

A COMPREHENSIVE REVIEW OF THE VESTIBULAR SYSTEM

Capstone Project

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By

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ABSTRACT

The vestibular system is housed within the bony labyrinth of the inner ear and is made up of three semicircular canals, which respond to angular head movements, and two otolithic organs, which respond to linear head movements. There are multiple diseases and disorders that can negatively impact the function of the vestibular system and result in symptoms including vertigo and imbalance. Audiologists perform many diagnostic evaluations to assist with the differential diagnosis of vestibular disorders, including videonystagmography (VNG), posturography, rotational chair, and vestibular evoked myogenic potentials (VEMP). These evaluations are essential in determining the site of lesion and treatment options for the dizzy patient.

The purpose of this capstone is to provide a comprehensive overview of the anatomy and physiology of the vestibular system, to review some common disorders that are often present in the dizzy patient, to summarize the evaluations that an audiologist may perform to assist in the differential diagnosis of a dizzy patient, and to present multiple case studies.

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LIST OF ABBREVIATIONS

BPPV	Benign Paroxysmal Positional Vertigo
CI	Cochlear Implant
CNS	Central Nervous System
CN VIII	Cranial Nerve VIII (Vestibulocochlear Nerve)
CT	Computed tomography
C-VEMP	Cervical Vestibular Evoked Myogenic Potential
dB	Decibel
DBN	Down Beat Nystagmus
ENG	Electronystagmography
IAC	Internal Auditory Canal
LBN	Left Beat Nystagmus
nHL	Normalized Hearing Level
RBN	Right Beat Nystagmus
SCC	Semicircular Canal
SCD	Superior Semicircular Canal Dehiscence
SCM	Sternocleidomastoid Muscle
SHA	Sinusoidal Harmonic Acceleration
SSRI	Selective Serotonin Reuptake Inhibitors
SVV	Subjective Visual Vertical
TRAPs	Trapezoidal Rotation
UBN	Up Beat Nystagmus
VCR	Vestibulo-Colic Reflex

VEMP	Vestibular Evoked Myogenic Potential
VNG	Videonystagmography
VOR	Vestibulo-Ocular Reflex
VRT	Vestibular Rehabilitation Therapy
VSR	Vestibulo-Spinal Reflex

CHAPTER 1

THE ANATOMY AND PHYSIOLOGY OF THE VESTIBULAR SYSTEM

Introduction

The balance system is comprised of three different portions: The vestibular portion of the inner ear, the visual system, and the somatosensory system, which combines the sense of touch and the ability for the body to sense its location in space (Fukuoka et al., 2001). These three systems work in unison to maintain balance for actions such as walking, running and dancing. If there is a malfunction within any one of these three systems, a person will experience anywhere from a slight disturbance such as unsteadiness while walking, to major disability like the inability to ambulate without aid from a cane.

The vestibular system is complex, and only a minority of audiologists work with the different evaluation methods that are within their scope of practice for the differential diagnosis of the dizzy patient. According to the American Academy of Audiology (2004), an audiologist “is uniquely qualified to provide a comprehensive array of professional services related to the ... identification, assessment, diagnosis, and treatment of persons with impairment of auditory and vestibular function”. The purpose of this capstone is to provide a comprehensive overview of the anatomy and physiology of the vestibular system, to review of some common disorders that are often present in the dizzy patient, to provide a summary of the evaluations that an audiologist may perform to assist in the differential diagnosis of a dizzy patient, and to present multiple interesting case studies of patients with vestibular dysfunction.

An Overview of the Anatomical Structures of the Vestibular System

I. The Bony and Membranous Labyrinth

The bony labyrinth, which is encased within the temporal bone, is comprised of the cochlea, the vestibule, and the semicircular canals (SCCs). The bony labyrinth is filled with perilymph, a fluid with a high sodium and low potassium composition. The chemical composition of perilymph is similar to the composition of cerebrospinal fluid, and perilymph communicates with cerebrospinal fluid through the cochlear aqueduct. The membranous labyrinth is suspended within the bony labyrinth by the perilymph and connective tissue. It is filled with endolymph, a fluid which is high in potassium and low in sodium (Hain & Helminski, 2007).

There are five distinct vestibular end organs that are contained within the membranous labyrinth of vestibular system: the three SCCs, named for their anatomical orientation in space (horizontal/lateral, superior/anterior, and inferior/posterior) and the two otolith organs, the utricle and the saccule (Hain & Helminski, 2007).

II. The Semicircular Canals

The three semicircular canals are essentially hollow tubes connected to the vestibule and filled with cochlear fluids that move freely within the canals in response to head movement. The purpose of the semicircular canals is to send communication to the brain regarding angular movements of the head within the anatomical plane on which they are located, for example, if a person shakes his head back and forth as if to signify the word “no”, the horizontal SCCs will be activated. The SCCs are positioned according to their anatomical name: the anterior and posterior SCCs intersect one another at 90 degree angles and the horizontal canal is tilted back 30 degrees from the horizontal plane. (Hain & Helminski, 2007). Each of the three SCCs end in a

widened bulb called the ampulla, which is attached to the vestibule. The ampulla contains the

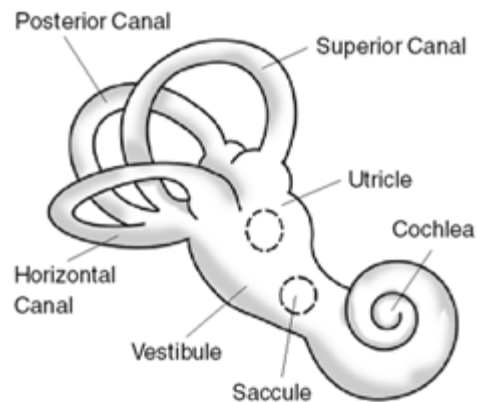


Figure 1. Illustration of the vestibular portion of the inner ear. Adapted from NIDCD, by the National Institutes of Health, 2003, retrieved from http://www.nidcd.nih.gov/health/balance/balance_disorders.asp. Copyright 2003 by the United States Department of Health and Human Services.

crista, which is a collective name for the cupula, the sensory cells of the vestibular system known as hair cells, and the ampullary crest. The ampullary crest is an elevated surface which makes up the floor of the ampulla; this is the location where the bases of the hair cells synapse with the vestibular nerve fibers. The

cupula is a gelatinous membrane that encompasses the hair cells of the vestibular system and separates them from the endolymph. The tallest stereocilia and the kinocilia of the hair cells within the SCCs project into

the cupula (Hain & Helminski, 2007).

III. The Otolith Organs

There are two otolith organs, known as the utricle and the saccule. These structures are located within the vestibule, with the saccule oriented

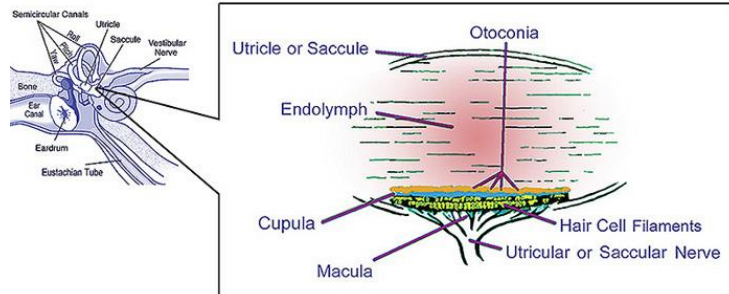


Figure 2. Illustration of the otolithic organs. Adapted from Wikipedia, by the United States Government, 2004, retrieved from <http://en.wikipedia.org/wiki/Otolith>. Copyright 2004 by the United States Government.

vertically and the utricle oriented horizontally. Similar to the SCCs, the otolithic organs also sense movement consistent with their orientation in space, meaning that the utricle senses linear movement in the horizontal

plane such as moving forward in a car, while the saccule senses linear movement in the vertical

plane such as going up in an elevator. Both structures are filled with endolymph (Schubert & Shepard, 2008).

The macula is the collective name for the sensory hair cells, the otolithic membrane, and the otoconia. This structure is located on the floor of the utricle and the medial wall of the saccule. The bases of the hair cells synapse with the vestibular nerve fibers through the floor of the utricle and the medial wall of the saccule. The stereocilia and the kinocilia of the hair cells are embedded in the otolithic membrane. The otoconia are calcium carbonate crystals that rest on the otolithic membrane. The hair cells in the saccule are oriented away from the striola, which is a dividing line down the middle of the otolithic organs, while the hair cells in the utricle are oriented towards the striola. The striola is curved, allowing for the otoliths to be sensitive to linear acceleration in multiple different directions (Shields & Gadre, 2004).

IV. *The Sensory Hair Cells of the Vestibular System*

There are two types of sensory hair cells located in the vestibular system: Type I and Type II, equal in number. The main differences between the two types of hair cells are their shape and the level of innervation. Type I hair cells are flask-shaped while Type II hair cells are cylindrical in shape. Type I hair cells receive more innervation than Type II hair cells; between one and five Type I hair cells are innervated by one vestibular nerve fiber while 30 to 40 Type II hair

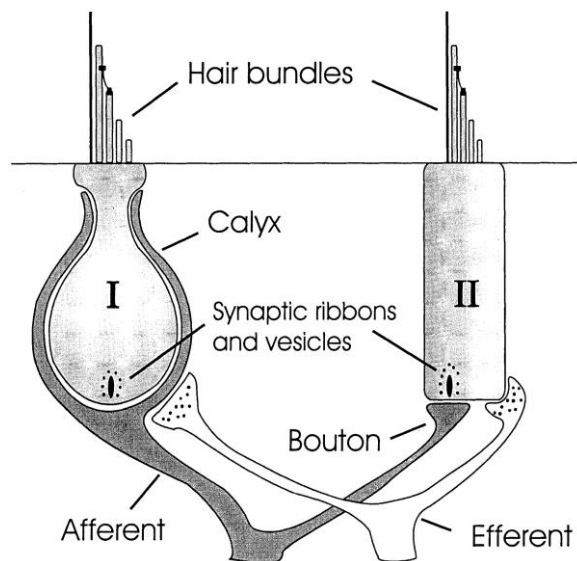


Figure 3. Type I and Type II Vestibular hair cells. Adapted from "Major potassium conductance in type I hair cells from rat semicircular canals: Characterization and modulation by nitric oxide", by J.W. Chen & R.A. Eatock, 2000, *Journal of Neurophysiology*, p. 140, Copyright 2000 by the American Physiological Society.

cells are innervated by one vestibular nerve fiber. The vestibular nerve fibers arise from Scarpa's Ganglion and send afferent communication from the vestibular end organs to the vestibular nuclei. From the vestibular nuclei, the signals are transferred to the extraocular nuclei to communicate with the ocular muscles (the Vestibulo-Ocular Reflex, or VOR), the spinal cord (the Vestibulo-Spinal Reflex, or (VSR) or Vestibulo-Collic Reflex, or (VCR)), or the cerebellum (Schubert & Shepard, 2008).

Vestibular hair cells are similar to auditory hair cells in multiple respects. The vestibular hair cells are topped with multiple stereocilia arranged according to height from shortest to tallest, similar to auditory hair cells. The vestibular hair cells differ from auditory hair cells because each vestibular hair cell has a single kinocilium, which is located next to the tallest stereocilia. In the semicircular canals, the kinocilia are all facing the same direction. In the anterior and posterior canals, the kinocilia are oriented away from the utricle, whereas in the horizontal canal the kinocilia point towards the utricle. Therefore, head movement to one direction or the other within the given plane causes all kinocilia to move in the same direction. In the otolithic organs, the kinocilia point away from the striola in the saccule and toward the striola in the utricle (Schubert & Shepard, 2008).

An Overview of the Physiology of the Vestibular System

I. The SCCs

As mentioned previously, the semicircular canals are hollow tubes that allow the endolymph to move freely within them. When a person moves his/her head at an angle, the endolymph within the SCC corresponding to the anatomical plane of head movement will move. This

movement causes the cupula to deflect, which causes deflection of the hair cells within the cupula. This deflection causes either hyperpolarization, which causes inhibition of an action potential, or depolarization, which causes excitation of an action potential, of the hair cells depending on the direction of the movement (Schubert & Shepard, 2008).

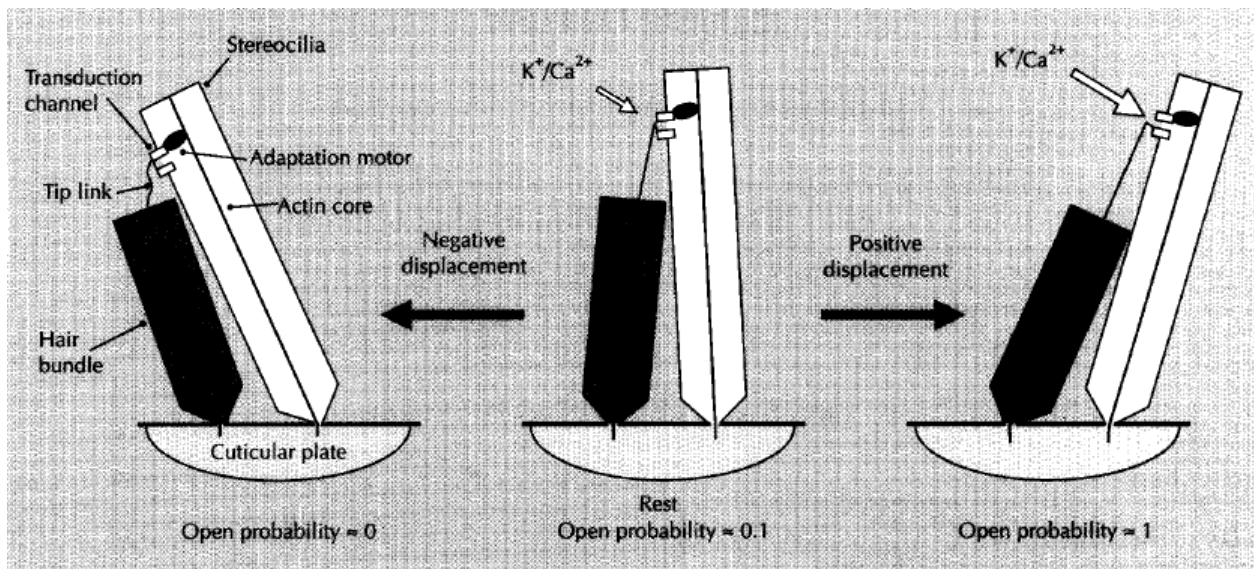


Figure 4. The displacement of hair cells, which leads to hyperpolarization or depolarization, allowing the transduction of mechanical stimuli into neural impulses. Depolarization occurs when the hair cell bends toward the kinocilium while hyperpolarization occurs if the hair cell bends away from the kinocilium. Adapted from “Molecular machinery of auditory and vestibular transduction”, by P.G. Gillespie, 1995, *Current Opinion in Neurobiology*, p. 450, Copyright 1995 by Current Biology.

Movement of the endolymph within the SCCs occurs in either an ampullopetal direction, or toward the ampulla, or an ampullofugal direction, or away from the ampulla. Ampullopetal flow evokes an excitatory response in the horizontal canal and an inhibitory response in the anterior and posterior canals. This occurs due to the organization of the kinocilium within the cupula, if the hair cell bends toward the kinocilium, depolarization occurs. If the hair cell bends away from the kinocilium, hyperpolarization occurs. The hair cells in the horizontal canal will excite when the endolymph moves in an ampullopetal motion while the hair cells in the posterior

and anterior canals will excite when the endolymph moves in an ampullofugal motion (Schubert & Shepard, 2008)

The SCCs are paired (the two horizontal canals are a pair; the right anterior and left posterior; and the left anterior and right posterior), which allows the brain to obtain the same information from two separate sources. The SCCs are paired as such due to the coplanar relationship, meaning that each pair is located on the same anatomical plane (Hain & Hillman, 1993), which allows for redundancy of the vestibular system. This permits central vestibular compensation, or a recovery of both gaze and postural stability (Curthoys & Halmagyi, 1995) to occur if one of the two pairs is damaged (Shields & Gadre, 2004).

II. The Otolithic Organs

Hyperpolarization or depolarization of the hair cells in the utricle and the saccule occur similarly to the semicircular canals, however the otolithic organs are sensitive to linear movement as opposed to angular movement. The tallest kinocilium of each hair cell is embedded into the otolithic membrane, which is topped by otoconia. The mass of the otoconia is greater than the mass of the endolymph, which causes it to be sensitive to changes in gravity and linear acceleration (Shields & Gadre, 2004). If displacement of the hair cell occurs toward the kinocilium, depolarization (excitation) occurs while if displacement occurs away from the kinocilium, hyperpolarization (inhibition) occurs (Schubert & Shepard, 2008).

III. Afferent Communication of the Vestibular System

Scarpa's Ganglion houses the cell bodies of the afferent vestibular nerve fibers and is located in the internal auditory canal (IAC) (Wright & Schwade, 2007). The vestibular nerve fibers, which arise from Scarpa's Ganglion, synapse at the bases of the vestibular hair cells and send afferent communication to the brain to be interpreted as movement. The vestibular nerve is

divided into two portions: the superior vestibular nerve, which innervates the anterior canal, horizontal canal, and utricle; and the inferior vestibular nerve, which innervates the posterior canal and saccule (Hain & Helminski, 2007). The resting firing rate for a healthy afferent vestibular system is often between 70 and 100 spikes per second. Excitation leads to an increase in the firing rate, while inhibition leads to a decrease in the firing rate (Schubert & Shepard, 2008). After the superior and the inferior portions of the vestibular nerve connect, these fibers join the auditory nerve while exiting the bony labyrinth and form Cranial Nerve VIII (CN VIII), the vestibulocochlear nerve. CN VIII travels through the IAC with the facial nerve. The afferent fibers of CN VIII terminate within the vestibular nucleus, which communicates with the cerebellum, spinal cord, extraocular nuclei, and the contralateral vestibular nucleus (Shields & Gadre, 2004).

IV. Efferent Communication- the Vestibulo-Ocular Reflex (VOR), Vestibulo-Spinal Reflex (VSR), and Vestibulo-Colic Reflex (VCR)

The central nervous system is constantly monitoring sensory inputs from the visual, vestibular, and proprioceptive systems and adjusting motor outputs, including eye movements and movements from the body, to maintain balance (Shields & Gadre, 2004). The influence of the vestibular system on the ocular system is called the VOR and the influence of the vestibular system on body position and compensatory movements to maintain balance is called the VCR and the VSR.

The purpose of the VOR is to allow the eyes to maintain clear sight of objects while the head is in motion. The fovea is the back of the eyeball, located in the center of the macula, and allows for sharp vision even while the head is in motion. A problem in the VOR may lead to retinal slip, or oscillopsia, a visual disturbance that may range from blurry vision to the perception of

stationary objects moving within the visual field (Bender, 1965). These problems are often exacerbated by movement. A healthy VOR will allow for eye movement that is equal in velocity

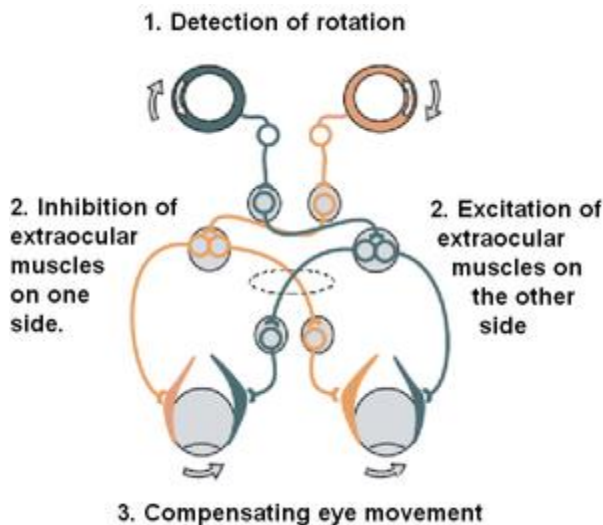


Figure 5. A simplified illustration of the VOR. Adapted from Wikipedia, by M. Haggstrom, 2007, retrieved from http://en.wikipedia.org/wiki/Vestibulo%20ocular_reflex. Copyright 2007 by Creative Commons Attribution.

and in the opposite direction of head movement.

Thus, the eye is able to remain fixated on an object while the head is in motion (Hain & Helminski, 2007). The visual system receives constant efferent communication from the vestibular system and constantly adjusts eye position based on head and body position. Eye

movement within the horizontal plane is controlled by two muscles: the lateral rectus, which moves the eye laterally or away from the nose, and the medial rectus which moves the eye medially or toward the

nose. Excitation of the horizontal semicircular canal leads to contraction of the ipsilateral medial rectus and contralateral lateral rectus, while inhibition leads to contraction of the contralateral medial rectus and the ipsilateral lateral rectus. Eye movement within the vertical plane is controlled by the superior rectus, which moves the eye in an upward direction, and the inferior rectus, which moves the eye in a downward direction. Torsional eye movements are controlled by the inferior and superior oblique muscles. Excitation of the posterior semicircular canal leads

to contraction of the ipsilateral superior oblique and the contralateral inferior rectus while inhibition causes contraction of the ipsilateral inferior oblique and the contralateral superior rectus. Excitation of the anterior SCC leads to contraction of the ipsilateral superior rectus and contralateral inferior oblique and inhibition causes contraction of the ipsilateral inferior rectus

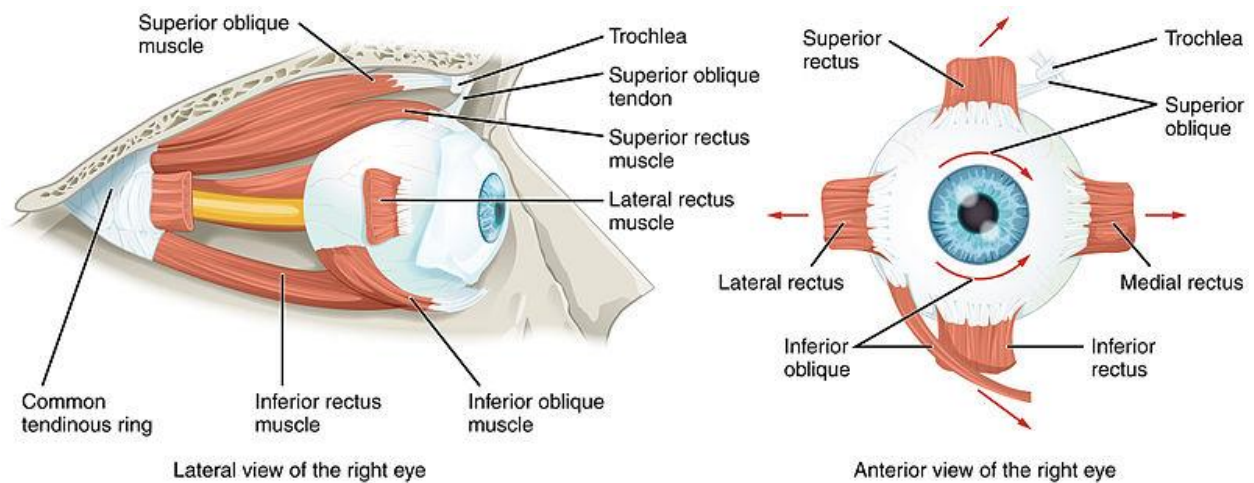


Figure 6. Ocular muscles of the right eye. Adapted from Extraocular Muscles, in *Wikipedia*, 2013, retrieved from http://commons.wikimedia.org/wiki/File:1412_Extraocular_Muscles.jpg. Copyright 2013 by Creative Commons Attribution.

and the contralateral superior oblique. This communication between the SCCs and the eye muscles allows for movements of the eyes to occur equal and opposite of head movement (Schubert & Shepard, 2008). Nystagmus may occur if the brain is receiving asymmetric input from the two ears. Nystagmus is an involuntary, repetitive eye movement which may occur in any plane that is controlled by the rectus or oblique muscles of the eyes- horizontal, vertical, or torsional (Straube et al., 2012).

The purpose of the VCR is to keep the head centered and stabilized over the neck while the body is in motion. The VCR receives efferent communication from the vestibular system regarding changes in position and communicates with the neck muscles, allowing them to react to these changes in head or body position in order to stabilize the head (Hain & Helminski, 2007). The purpose of the VSR is to generate body movements to maintain balance and prevent

falls. The VSR includes the spine and ankles, structures that are responsible for maintaining postural stability (Schubert & Shepard, 2008). Audiologists have a greater focus on the VOR system than the VSR and VCR systems during evaluations, as the VOR plays a crucial role during videonystagmography (VNG), rotational chair, and posturography evaluation while the VSR and VCR are evaluated during C-VEMP and posturography evaluations. The ability to examine the VOR system enables the audiologist to find localizing information regarding peripheral vestibular disorders (Schubert & Shepard, 2008).

CHAPTER 2

COMMON DISORDERS OF THE VESTIBULAR SYSTEM

There are multiple disorders that can impact various portions of the vestibular system, including the semicircular canals, the otolith organs, the vestibular nerve, or the central vestibular system. The following section discusses some of the more common vestibular disorders; however it is not a comprehensive list. The purpose of this chapter is to discuss some common vestibular disorders, different causes of the disorders, symptomology, and treatment options. Audiologists are able to aid in determination of the differential diagnosis between many of these disorders with a comprehensive vestibular evaluation.

Meniere's Disease

I. Incidence, Definition and Cause of the Disorder

Meniere's disease is a chronic disorder with no known cure which affects approximately 15 out of 100,000 individuals per year. It impacts one ear more commonly, however approximately 24% of patients are diagnosed with Meniere's disease bilaterally. When the second ear has a delayed onset in comparison to the first ear, the disease presents an average of 7.6 years in the second ear following the first ear (House et al., 2006). Approximately 10% of patients with Meniere's disease have a positive family history of the disease (Fung et al., 2002). Initial onset usually occurs in the fourth to sixth decade of life (Clemmens & Ruckenstein, 2012).

Meniere's disease is historically thought to be caused by endolymphatic hydrops, or an abnormal amount of endolymph within the inner ear. Endolymphatic hydrops may occur as a result of trauma, an inflammatory disease, or a disturbance in the process of secreting and

absorbing endolymph within the inner ear (Schuknecht, 1976). Some studies suggest that Meniere's disease may be caused by an autoimmune disorder (Greco et al., 2012), however this section focuses on Meniere's disease which is thought to be a result of endolymphatic hydrops.

Temporal bone studies have demonstrated that not all cases of endolymphatic hydrops resulted in the typical symptoms of Meniere's disease (House et al., 2006). The increase of endolymph causes the basilar membrane and Reissner's membrane to stretch. The excess fluid and pressure build-up puts unnecessary stress on the auditory and vestibular hair cells and causes hair cell damage and eventually hair cell death, and with this damage the classic symptoms associated with Meniere's disease occur (Baloh & Halmagyi, 1996).

II. *Symptomology*

There are four classic symptoms that are commonly reported in Meniere's disease- the "triad" of Meniere's disease is tinnitus, hearing loss, and dizziness; while a fourth commonly reported symptom is aural fullness (Semaan et al., 2005). An attack of Meniere's disease usually occurs with little or no warning and there are few known provoking causes of the attacks.

One characteristic symptom of Meniere's disease is fluctuating sensorineural hearing loss, beginning in the low frequencies and eventually impacting the high frequencies as well. Audiometric configuration varies based on the stage of Meniere's disease, and over time, the hearing stops fluctuating and stabilizes to a permanent hearing loss with audiometric configuration changing from a rising to a flat loss (Stahle, 1984, Kotimaki et al., 2003). Tinnitus, which is often referred to as a low-frequency "roaring" sound, and aural fullness are two more common symptoms that occur in fluctuations as well. The patient may also experience non-stop vertiginous episodes, which occur without warning and may last anywhere from 20 minutes to 24 hours, but for most people the attacks last for 2 to 3 hours. After the vertigo subsides, the

patient may experience imbalance, nausea or exhaustion. Some patients may experience aural fullness prior to the attacks, while others do not (Baloh & Halmagyi, 1996).

III. Treatment of the Disorder

While Meniere's disease does not have a cure, there are multiple methods of management. The goals in the management options are to keep the bodily fluids stabilized to avoid fluctuations in endolymph. This includes controlling sodium intake and the use of diuretics. The patient may also be prescribed vestibular suppressants, such as anticholinergics, benzodiazepines, or antihistamines, to help stabilize them during the attacks (Baloh & Halmagyi, 1996). Intratympanic or oral steroids may be prescribed during fluctuations in hearing to help return hearing to the baseline. The steroids are beneficial for two reasons: They decrease inflammation within the inner ear and promote blood flow. In severe cases of vertigo, vestibulotoxic medications such as gentamicin may be used to destroy the vestibular function completely in the impacted ear (Hain, 2013).

Surgical treatment of Meniere's disease is only utilized in very severe cases where other management strategies have not been successful. Endolymphatic sac decompression is a surgical option to relieve pressure in the inner ear. This surgery may include the insertion of a shunt to direct excess endolymph flow out of the inner ear (Ostrowski & Kartush, 2003). A labyrinthectomy, or removal of the vestibular portion of the inner ear, may also be considered in severe cases of vertigo (Brinson et al., 2007). Another surgical option in Meniere's disease is a cochleosacculotomy, which involves the creation of a fistula in the sacculle between the endolymph and the perilymph to allow for relief of pressure differences between the two areas (Baloh & Halmagyi, 1996). Complications of these surgeries are rare and these procedures are considered to be relatively safe (Brinson et al., 2007). Surgical or vestibulotoxic methods of

destroying the vestibular function of the impacted ear allow the patient to centrally compensate because they will no longer experience fluctuations in vestibular function. Approximately 96% of patients with Meneire's disease who underwent a vestibular nerve section as treatment experienced relief of vertigo, while 75% of patients who underwent gentamicin treatment experienced relief (Telian & Wiet, 2008).

Vestibular Neuronitis and Labyrinthitis

I. Definitions and Causes of the Disorders

Vestibular neuronitis is a disorder which occurs due to viral inflammation of the superior portion of the vestibular nerve (Hain, 2012). One common viral infection that may lead to vestibular neuronitis is the Herpes virus (Baloh et al., 1996). Labyrinthitis, however, is often caused by a viral or bacterial infection such as an upper respiratory infection (Stokroos et al., 1998), meningitis or otitis media (Bennett, 2008). The annual incidence of vestibular neuronitis and labyrinthitis in the United States is approximately 150,000 patients (Marill et al., 2013) and approximately 5% of all cases of dizziness are thought to be caused by one of these two disorders (Hain, 2012).

II. Symptomology

One symptom experienced by patients with vestibular neuronitis is the sudden onset of acute vertigo, which may last as long as several days and may be accompanied by nausea, vomiting, and impaired vision (Shupak et al., 2008). The patient may also experience disequilibrium or imbalance for weeks after onset of initial vertigo, although subsequent episodes of true vertigo are unlikely. A patient will not experience associated auditory symptoms such as hearing loss or

tinnitus in the case of vestibular neuronitis, which is one method of distinguishing this disorder from labyrinthitis (Bennett, 2008).

The symptoms of labyrinthitis are very similar to the symptoms of neuronitis, including acute vertigo, nausea, vomiting, and lasting imbalance. Unlike neuronitis, labyrinthitis is often accompanied by associated auditory factors such as high frequency, unilateral sensorineural hearing loss and high frequency tinnitus. The hearing loss may be diagnosed by audiologic evaluation and patients may report the tinnitus during case history. These auditory symptoms may be temporary or permanent (Hain, 2012) and audiologic monitoring is crucial to monitor the progression of these symptoms.

III. Treatments of the Disorders

Vestibular neuronitis is often treated with steroids (Shupak et al., 2008), vestibular suppressants such as anticholinergics, benzodiazepines, or antihistamines, or vestibular rehabilitation therapy (VRT) (Walker, 2009). Treatment for labyrinthitis is similar, including the use of steroids, vestibular suppressants, and VRT (Hain, 2012). VRT includes exercises that aid in adaptation or habituation for a vestibular dysfunction and may include balance, walking, endurance, or strengthening exercises depending on the individual needs of the patient (Boyer et al., 2008).

Benign Paroxysmal Positional Vertigo (BPPV)

I. Definitions and Causes of the Disorder

Benign Paroxysmal Positional Vertigo (BPPV) is the most common inner ear disorder that causes dizziness. Approximately 2.4% of the population has experienced BPPV at some point in their lives (von Brevern et al., 2007). BPPV occurs when the otoconia come loose from the

otolithic membrane of the utricle and fall into any one of the three SCCs, causing it to become sensitive to changes in gravity. Posterior canal BPPV is the most common due to the anatomical location of the posterior canal in relation to the utricle (Bennett, 2008). The cause of BPPV is most often idiopathic; however it may also be caused by head trauma, whiplash, or as the result of surgery of the ear or dental surgery (Chiarella et al., 2008).

There are two types of BPPV- cupulolithiasis and canalithiasis. Cupulolithiasis describes particles that are attached to the cupula of the semicircular canal while canalithiasis describes free-floating otoconia within the SCC. Canalithiasis is more common than cupulolithiasis (Bittar et al., 2011) and cupulolithiasis is seen in less than 5% of cases of BPPV (Miles, 2012). Nystagmus caused by canalithiasis lasts between 5 and 30 seconds and then fatigues. When the patient is moved from the supine to the sitting position, the nystagmus reverses direction (Bittar et al., 2011). Nystagmus caused by cupulolithiasis does not fatigue and it will reverse directions when the impacted canal is moved 180 degrees to the opposite direction (Miles, 2012).

II. Symptomology

Patients with BPPV often report sudden and severe attacks of vertigo which are provoked by movement in a certain direction, most commonly when turning over in bed or getting into or out of bed. The attacks last for fewer than 30 seconds in canalithiasis; or for as long as the patient's head is in the provoking position in cupulolithiasis. Some patients also report nausea, lightheadedness or disequilibrium following the episode for up to 24 hours (Parnes et al., 2003).

III. Treatments of the Disorder

BPPV often resolves on its own within 6 months with no treatment. If treatment is required, however, a canalith repositioning maneuver such as the Epley maneuver may be utilized to reposition the otoconia to their appropriate position on the otolithic membrane (Parnes et al.,

2003). Very rarely, surgical intervention is considered. This surgery, posterior semicircular canal occlusion, requires that the posterior semicircular canal is plugged with either a fascia graft or muscle from the patient to prevent the otoconia from falling back into the canal (Baloh & Halmagyi, 1996; Shaia et al., 2006).

Superior Canal Dehiscence (SCD)

I. Definitions and Causes of the Disorder

Superior Canal Dehiscence (SCD) occurs when an individual has an absence or thinning of a section of bone in the superior semicircular canal. SCD may be caused by head trauma, a complication during surgery, an issue during embryological development, or in cases where the bone is already thin, increased pressure within the head during coughing, sneezing, during the Valsalva maneuver, or straining as in to lift a heavy object (Telian & Wiet, 2008). SCD causes a “third window effect”. In a normally functioning ear, the stapes compresses cochlear fluids at the oval window and this pressure is relieved at the round window. The third window changes the pressure gradient within the system. The prevalence of SCD among the general population is approximately 0.5% (ASHA, 2014).

II. Symptomology

Common symptoms of SCD include Tullio’s phenomenon, or vertigo induced by sound, Hennebert’s sign, or vertigo that is induced by pressure changes, low frequency conductive hearing loss, pulsatile tinnitus, aural fullness, chronic imbalance or disequilibrium (Hillman et al., 2006), and autophony, or the sensation of hearing body noises, such as breathing or the heartbeat, or the person’s own voice, at an unusually or uncomfortably loud level (Banerjee et al., 2005). The low frequency conductive hearing loss occurs because the “third window”

changes the impedance of the cochlea and does not allow for adequate transmission of low-frequency sounds while allowing for increased transmission of bone-conducted stimuli (McEvoy et al., 2013). During the vestibular evaluation, the audiologist may see vertical nystagmus because the superior canal is the impacted canal (Telian & Wiet, 2008).

III. Treatments of the Disorder

SCD may be treated by avoidance of provoking factors such as loud sounds or pressure changes (Banerjee et al., 2005) or surgical repair of the dehiscence (Hillman et al., 2006). Surgical treatment should be reserved for very severe cases due to the risk of surgical complications (Brantberg et al., 2001). Amplification may be considered when warranted by the degree of hearing loss; however Tullio's phenomenon must be considered when the hearing aids are being set (ASHA, 2014). The audiologist must consider the maximum power output of the hearing aid and ensure that the hearing aid is not amplifying to a level which will provoke dizziness in the patient.

Perilymphatic Fistula

I. Definitions and Causes of the Disorder

Perilymphatic fistulas occur when perilymph leaks from the oval or round window or from the bony labyrinth and most often occur unilaterally (Hain, 2012). Some fistulas are congenital in nature, for example if a Mondini malformation of the inner ear is present, while others are acquired, for example due to head trauma, surgical trauma, or changes in intracranial or atmospheric pressure (Bennett, 2008). Perilymphatic fistulas are rare and repair of this disorder accounts for approximately 0.2% of otologic surgeries (Muntarhorn & Webber, 1987).

II. Symptomology

Symptoms of perilymphatic fistulas include vertigo which occurs at the time of the fistula and lasts for seconds or minutes, imbalance which may last for months, tinnitus, aural fullness, unilateral sensorineural hearing loss, and nausea (Kimitsuki et al, 2003). Patients may also feel dizzy when experiencing pressure changes, such as when sneezing, or straining such as while lifting heavy objects (Castillo & Roland, 2007).

III. Treatments of the Disorder

Treatment of a perilymphatic fistula may include bed rest or reduction of physical activity to allow the fistula to heal itself, the use of a pressure equalization (PE) tube to help avoid barotrauma during pressure changes in cases of a dysfunctional Eustachian tube (Hain, 2013), VRT, or surgical repair of the fistula (Fitzgerald, 1995).

Migraine-Related Vertigo

I. Definitions and Causes of the Disorder

Vestibular migraines are defined as attacks of vertigo that are caused by migraines or occur in conjunction with migraines (Lempert & Neuhauser, 2009). A migraine is described as a “disabling, unilateral, recurrent, intermittent and pulsating headache that is associated with nausea and sensitivity to light and sound” (International Headache Society, 2014). Over 10% of people worldwide experience migraines (Migraine Research Foundation, 2014); approximately 1% of people have vestibular migraines (Bussone, 2003) and the incidence of migraines is three times higher in people who have vertigo (Fotuhi et al., 2009).

II. Symptomology

There are multiple symptoms that individuals with vestibular migraines may experience, including spontaneous or positional vertigo which may last anywhere from seconds to days, migraines with or without aura, photophobia, or sensitivity to light, and phonophobia, or sensitivity to sound. Patients may also experience motion sensitivity or nausea induced by head movements (Lempert & Neuhauser, 2009).

III. Treatments of the Disorder

Treatment options for vestibular migraines include medication to treat the symptoms, a balanced diet, exercise, and a regular sleeping schedule (Fotuhi et al., 2009). Medications may include antidepressants, propranolol, or meclizine. Patients may also avoid any triggers for their migraines (ex: stress) or undergo vestibular rehabilitation (Lempert & Neuhauser, 2009).

Miscellaneous Central Vestibular Disorders

I. Definitions and Causes of the Disorders

There are multiple causes of dizziness that are considered central in nature, including cerebellar infarction (Dieterich, 2007), medullary infarction (Choi & Kim, 2009), central neurodegenerative diseases such as Parkinson's disease (Pan et al., 2008) or multiple sclerosis (Ramadan, 2008), or acoustic neuroma (Cohen et al., 2002). Vestibular migraines are also a central cause of dizziness; however this disorder is discussed in the previous section. Patients may also present with psychological dizziness if they have anxiety or present with panic attacks or hyperventilation. Hyperventilation is considered one of the most common causes of dizziness in the young population (Bennett, 2008).

II. Symptomology

Symptoms of central vestibular disorders may vary based on the site of lesion, and the patient may have difficulty providing the clinician with an exact time of onset. Patients with central vestibular disorders may experience slow, constant subjective vertigo and imbalance while standing and walking. They may also complain of lightheadedness (Shepard, 2009). Patients with a central nervous system disorder may present with ataxia, which is unsteadiness and clumsiness while walking (Bennett, 2008).

III. Treatments of the Disorder

Treatments of central vestibular disorders vary widely depending on the cause of the disorder. Some treatments include medical management, for example resection of an acoustic neuroma (Cohen et al., 2002). In the case of a neurodegenerative disease such as multiple sclerosis, treatment may include immunotherapy treatment or medications to treat the patient's symptoms (Ramadan, 2008). Patients with psychological causes of dizziness may be treated with medications such as selective serotonin reuptake inhibitors (SSRIs) or varying types of psychotherapies (Staab, 2008).

CHAPTER THREE

DIFFERENTIAL DIAGNOSIS

Audiologists play a significant role in the differential diagnosis of balance disorders. A comprehensive assessment consisting of a full audiometric evaluation, vestibular evoked myogenic potentials (VEMP), rotational chair, posturography, and videonystagmography (VNG) often leads to the diagnosis and helps to determine the course of management for a patient. Patients with vestibular disorders often require management from multiple disciplines, including but not limited to audiology, neurology, physical therapy, and otology. Audiologists must make appropriate referrals to the different specialties based on the test results to help ensure proper management and patient care.

Case History

The case history is an essential part of the differential diagnosis of a dizzy patient. The umbrella term of “dizziness” can describe a multitude of different symptoms, from vertigo to imbalance to lightheadedness. Dizziness ranks third on the list of most common complaints seen in hospitals and clinics (Bennett, 2008). Different vestibular disorders present with different symptoms and a thorough case history will help guide the audiologist to the proper diagnosis.

The clinician must first differentiate what type of dizziness the patient is experiencing. It is crucial to determine if the patient has experienced true vertigo as a symptom because vertigo is often related to disorders of the inner ear while patients who experience other types of dizziness may have a disorder of the cardiovascular system, the central system, the ocular system, or a systemic disease (Bennett, 2008). The clinician must also determine how long the dizziness lasts once it is provoked. The time course of the dizziness is another important aspect for the

differential diagnosis. Some vestibular disorders present with dizziness lasting a few seconds or minutes, such as BPPV, SCD, or perilymphatic fistulas. Other disorders present with dizziness lasting several minutes to several hours, such as Meniere's disease, migraines, or labyrinthitis. The audiologist must also determine how many attacks of dizziness the patient experiences. A patient who has experienced only one episode of dizziness may have labyrinthitis or neuronitis, while patients who have experienced multiple episodes may have Meniere's disease, BPPV, or migraines (Bennett, 2008).

The case history is an important time to determine what triggers the patient's symptoms. Triggers are another important piece of information when determining what may be the cause of the dizziness. For example, dizziness provoked by movement may be indicative of BPPV while dizziness provoked by loud sounds may be indicative of SCD. Patients whose symptoms are provoked by stress may have a form of psychological dizziness. The audiologist should also ask the patient to describe any symptoms they experience in conjunction with the dizziness. A patient who also experiences hearing loss with the dizziness may have Meniere's disease or labyrinthitis, while a patient who experiences phonophobia may have vestibular migraines (Bennett, 2008).

It is important for the audiologist to determine if the patient has any additional significant problems in their medical history during the case history as well. Multiple diseases and disorders may have dizziness as a symptom, including a history of seizures, migraines, neurological disorders, hypertension or high blood pressure, or psychiatric diseases such as anxiety (Bennett, 2008). The audiologist must also determine if the patient is taking any medications that may include dizziness as a side-effect, as approximately 25% of medications do. Some common medications that include dizziness as a side-effect are anti-anxiety and anti-depression

medications, diuretics, chemotherapy medications, antibiotics, and anti-inflammatory medications (Bennett, 2008). A thorough case history will help guide the clinician through the remainder of the appointment.

Audiologic Evaluation

I. Tympanometry

Tympanometry measures the mobility of the tympanic membrane and ossicles within the middle ear by varying the pressure in the ear canal (deBonis & Donohue, 2008). It is important to perform tympanometry, along with otoscopy, to determine the health of the ear canal and middle ear system prior to many vestibular evaluations. Bithermal caloric evaluations with water cannot be performed if a patient has a perforated tympanic membrane. A patient with a perforated tympanic membrane may demonstrate a hyperactive response on the ipsilateral side if bithermal caloric testing is performed with air due to an increase of temperature transduction (Spector, 1967). It is also important to rule out conductive hearing losses through both tympanometry and pure-tone audiometry for accurate C-VEMP and bithermal caloric results to be obtained (Seo et al., 2008).

As mentioned previously, patients with a perilymphatic fistula may experience dizziness with changes in pressure, for example during a sneeze or while lifting heavy objects. A “fistula test” may be performed where a perilymphatic fistula is suspected by varying the pressure within the ear canal with an immittance bridge. A fistula test is considered to be positive when changes in pressure in the ear canal evoke subjective dizziness in the patient. Nystagmus may be noted during this test as well (Causse et al., 1983).

II. Acoustic Reflexes

Acoustic reflexes are measured when a high-intensity sound stimulus is delivered into the ear and causes the stapedius muscle to contract, which leads to a stiffening of the ossicular chain and tympanic membrane. Any disruption of sound transmission, such as fluid behind the tympanic membrane or dislocated ossicles, may lead to an elevated or absent acoustic reflex. Acoustic reflexes may be measured either through the ipsilateral or contralateral ear (Clark et al., 2007).

Acoustic reflexes may be helpful in the diagnosis of retrocochlear pathology. As mentioned previously, patients with a tumor on CN VIII may experience dizziness as a symptom. The audiologist may note a CN VIII pattern during acoustic reflexes, which may be suggestive of an acoustic neuroma. A CNVIII pattern will result in absent ipsilateral reflexes on the side of the lesion and absent contralateral reflexes because the sound transmission is interrupted by the tumor on the nerve (Clark et al., 2007).

III. Pure-Tone Audiometry

The auditory and vestibular systems are very closely related, as they are housed in the same bony labyrinth, share the same fluids (endolymph and perilymph), and both communicate with the brain via CN VIII. For this reason, it is essential to test hearing function any time a patient complains of balance issues.

Many vestibular dysfunctions have a characteristic audiometric configuration which will aid in differential diagnosis. For example, patients with Meniere's disease often present with a low-frequency fluctuating sensorineural hearing loss (Stahle, 1984) while patients with SCD often have a hearing loss that is low frequency and conductive in nature (Hillman et al., 2006). Patients with an acoustic neuroma may present with an asymmetric sensorineural hearing loss (Suzuki et al., 2010).

Videonystagmography (VNG)/Electronystagmography (ENG)

I. Explanation of the Test

The videonystagmography (VNG) or electronystagmography (ENG) evaluation is considered the “gold standard” for determining if an inner ear disorder is the cause of the patient’s dizziness, because of its ability to gain ear-specific information (Hain, 2012), and the ability to determine the state of compensation of the lesion. A physician may choose to order a VNG/ENG if a peripheral or central vestibular lesion is expected.

In a VNG evaluation, the patient wears goggles that are equipped with infrared cameras within the lenses that allow the clinician to visualize the patient’s eyes throughout testing. The cameras lock onto the patient’s pupils and record eye movements, measuring any nystagmus that may be present. With an ENG system, surface electrodes are used to measure the corneoretinal potentials and produce tracings of nystagmus. Nystagmus may be horizontal or vertical. Right beat nystagmus (RBN) and left beat nystagmus (LBN) are horizontal forms of nystagmus and up beat nystagmus (UBN) and downbeat nystagmus (DBN) are vertical forms of nystagmus.

There are four subtests within the VNG/ENG: oculomotor tests, positional and positioning tests, horizontal head shake, and bithermal caloric evaluation. Oculomotor evaluation includes saccades, or rapid eye movements used to locate and focus on an object; smooth pursuit, or maintaining a stable gaze on a moving object; optokinetics (OPKs), or watching objects move within the visual field; and gaze testing, or the ability to maintain a stable gaze in the center, rightward, leftward, upward, or downward positions. Patients who demonstrate abnormalities during saccades, smooth pursuit, or OPKs likely have a disorder in the central nervous system (Shepard & Schubert, 2008).

Gaze testing may be used to evaluate for either central or peripheral vestibular lesions. Gaze testing may be performed both with vision enabled and vision denied. If nystagmus is noted in the vision denied condition and does not suppress or increases in intensity with fixation, it is considered a central indicator. If the nystagmus does suppress, it is an indicator of peripheral pathology (Shepard & Schubert, 2008). Spontaneous nystagmus that occurs during center gaze should be noted because it will impact interpretation throughout the remainder of the testing. Spontaneous nystagmus is often the result of acute (non-compensated) asymmetry of vestibular input from the ears (Hegemann et al., 2007).

The patient must be mentally alert throughout any test that is performed with vision denied or during bithermal caloric evaluation to reduce central suppression of nystagmus (McGovern & Fitzgerald, 2008). The response from the VOR may be reduced if the patient is not mentally alert throughout testing. The clinician may ensure that the patient is mentally alert throughout the evaluation by having the patient participate in active tasking activities, such as answering simple math questions or responding to reflexive questions, such as “How old are you?” (Davis & Mann, 1987).

Positioning evaluations includes the Dix-Hallpike, while positional evaluations includes the supine, head and body right, and head and body left positions. During the Dix-Hallpike, the patient is seated upright and the head is turned to either the right or the left at a 45 degree angle. The patient is rapidly brought to a supine position with the head extended approximately 20 degrees backward off of the examination table. This position allows the posterior canal to be positioned lateral to the floor. The patient remains in this position for approximately 30 seconds while the clinician evaluates for the presence of any nystagmus or subjective vertigo. The patient

is then returned to a seated position and the test is repeated on the other side. The Dix-Hallpike is used to evaluate for both posterior and anterior canal BPPV.

During positional testing, the clinician is evaluating if the vestibular system is responding symmetrically to changes in head position. The patient is moved to a supine position, the head is turned to the left and to the right, and the patient lies on the right and left side of their body. The clinician evaluates for nystagmus in any of these static positions. If nystagmus is observed, the evaluation is repeated in that position with vision enabled.

The high-frequency headshake involves the patient seated upright and with the head tilted downward at a 30 degree angle in order to position the horizontal SCC lateral to the floor. The clinician actively moves the head back and forth in a horizontal motion for 20 cycles at a frequency of approximately 2 Hz. The patient remains in this position with the eyes open and the clinician evaluates for any nystagmus immediately post-headshake. The high-frequency headshake is used to evaluate for both central and peripheral lesions.

Bithermal caloric evaluation is utilized to measure the symmetry of the peripheral vestibular system while the horizontal SCCs are hyperpolarized and depolarized, simulating a low-frequency head turn. The patient is moved to a supine position with the head tilted forward 30 degrees, allowing for the horizontal SCCs to be positioned lateral to the ground. Each ear is irrigated with both a cool and a warm stimulus, either air or water depending on the system being utilized, for a total of four irrigations lasting 30 seconds each. The irrigation causes a temperature change in the endolymph within the horizontal SCC, which changes its density. The change in density causes the endolymph to move within the canal, which in turn causes the cupula to deflect and results in an excitatory or inhibitory response from the hair cells. After the irrigation is complete, the patient must remain mentally alert to avoid central suppression of the

nystagmus (McGovern & Fitzgerald, 2008). The nystagmus is recorded for approximately one minute following the irrigation; following this the patient must fixate on a stimulus. Fixation will cause a marked reduction in the nystagmus in a normally functioning individual.

II. Anatomy Involved

The VNG/ENG will give the clinician insight regarding the health of the horizontal and posterior SCCs, CN VIII, the cerebellum, the brainstem, and the VOR pathway.

III. Normal and abnormal findings

Oculomotor tests involve tracking a target with the eyes and includes saccades, smooth pursuit and OPKs. Normal saccades occur when a person is able to track a target with their eyes within a normal latency and accuracy based on normative data. It is considered abnormal for a patient to have a prolonged latency or to demonstrate hypometria, (undershooting the target) or hypermetria (overshooting the target). A glissade is also considered abnormal, and occurs when the patient's eyes drift from the target. Smooth pursuit is considered normal when the patient is able to track a moving target with a stable gaze in regards to gain and symmetry. Abnormal results are noted when the score is asymmetric, or if the patient can follow the target in one direction but struggles to follow it in the opposite direction. It is also considered abnormal for the patient to demonstrate catch-up saccades while tracking the target. OPKs are considered normal when the patient demonstrates symmetric nystagmus during the task. It is considered abnormal if the patient is unable to complete the task or the response is not symmetric. Abnormal findings during oculomotor tests are often considered to be a central indicator, however it is considered a peripheral indicator if nystagmus is noted during smooth pursuit (Shepard & Schubert, 2008).

Gaze testing is often performed with vision denied first, and if the patient demonstrates nystagmus it is repeated with vision enabled. No nystagmus should be noted during gaze testing

in a patient with a normal vestibular system. If the patient demonstrates nystagmus in the vision denied condition, but it suppresses with vision enabled, it is considered a peripheral finding. If the nystagmus does not suppress with fixation, it is considered a central finding (Shepard & Schubert, 2008).

During positioning evaluations, a patient with a normal peripheral and central vestibular system will not have nystagmus following the high-frequency head shake or the Dix-Hallpike maneuver. If nystagmus is noted post-headshake and it beats in a horizontal manner toward the healthy ear, it is a peripheral indicator. If the nystagmus has a vertical component, is prolonged, or is disconjugate, it is considered a central indicator (McCaslin et al., 2008). Patients with BPPV will often demonstrate nystagmus during the Dix-Hallpike. For example, if a patient has posterior canal BPPV, they will demonstrate geotropic rotary nystagmus, or torsional nystagmus which beats toward the ground, in the head hanging position that changes direction when returned to the seated position when the head is turned toward the impacted ear (Bittar et al., 2011).

A patient with a normal vestibular system will not demonstrate nystagmus in any position during positional testing. If a patient does demonstrate nystagmus, it may be indicative of central or peripheral vestibular dysfunction (Brandt, 1997). Geotropic nystagmus or nystagmus that is transient in nature is most often suggestive of a peripheral lesion, while ageotropic, vertical or persistent nystagmus may be suggestive of a central lesion or pharmacological involvement (Gans & Yellin, 2007).

A patient with a normal peripheral vestibular system will demonstrate symmetric and robust vestibular responses during bithermal caloric evaluation. A patient with a peripheral vestibular lesion may demonstrate a unilateral weakness, which may be the result of either a

hyperfunction or hypofunction of one horizontal SCC. If the patient demonstrates a unilateral weakness, however no spontaneous nystagmus is noted during gaze testing, the lesion has likely compensated (Yamamoto et al., 2000).

IV. Limitations of the Test and Contraindications to Performing the Test

A limitation of the ENG is that the clinician cannot visualize the eyes throughout the evaluation and eye movements cannot be recorded for later review. In addition, torsional nystagmus cannot be recorded. Also, electrical noise or artifact from muscle movements may interfere with the ENG recording (Gananca et al., 2010). For these reasons, the VNG is preferable to the ENG.

On the other hand, the VNG equipment is more expensive than the ENG equipment, and recordings may be noisy in patients with ptosis or excessive blinking. The eyes must remain open during the VNG, while nystagmus may be recorded with the eyes closed with the ENG. The camera in the VNG equipment may have difficulty locking onto the pupil if the patient has a disease of the eye (Gananca et al., 2010).

Modifications may be required during the Dix-Hallpike for patients with neck or back issues. During the modified Dix-Hallpike, the audiologist will stand behind the patient and provide support with their hands to the patient's neck and back. The examiner must ensure full support of the patient's neck while the neck is hyperextended and the head is hanging off of the examination table. If the Dix-Hallpike cannot be performed because the patient is unable to hyperextend their neck, the side-lying maneuver may be used instead. In this maneuver, the patient begins in a seated position with the head turned 45 degrees to the left or to the right (the head is turned to the right if the left ear is being evaluated and vice versa). The patient then lays to the side with the head looking up to the ceiling and the legs are brought up to the examination

table. In this position, the patient's head and neck are supported entirely by the table (Roberts & Gans, 2008).

V. Recommendations based on test results

The audiologist may refer the patient to neurology if a central lesion is suspected or refer to otology if a peripheral lesion is suspected. The audiologist may also refer the patient for VRT if the lesion is uncompensated to assist with central compensation (Boyer et al., 2008). If BPPV is suspected, the audiologist may perform the Epley maneuver to reposition the otoconia. . The Epley maneuver is a canalith repositioning maneuver which involves a series of head and body positions which allow gravity to act on the otoconia, which are free-floating in the posterior semicircular canals, to move into the utricle (Richard et al., 2005).

Rotational Chair

I. Explanation of the Test

Rotational chair testing is considered to be the “gold standard” in the evaluation of bilateral vestibular lesions, such as patients with bilateral Meniere's disease (Gillespie & Minor, 1999). It is also utilized for patients who are currently taking vestibulotoxic medications, such as gentamicin or aminoglycosides, to monitor the vestibulotoxic effects of the medication on the patient's vestibular system (Gillespie & Minor, 1999). There are three main components of rotational chair evaluation: Sinusoidal harmonic acceleration (SHA), trapezoidal rotation (TRAPs), and static and dynamic subjective visual vertical (SVV). All testing is completed with the patient secured in a motorized chair that undergoes various movements. Testing is completed in darkness in order to avoid fixation on a visual stimulus (Brey et al., 2008).

SHA evaluates the VOR as the patient is moved in a horizontal motion back-and-forth. During SHA, the patient is rotated alternately to the left and right at varying frequencies from low to high. The gain, phase, and symmetry of eye movements are analyzed during this evaluation. During TRAPs, the patient is rotated at a constant velocity for sixty seconds and subsequently stopped abruptly. The period of time that is required for the nystagmus to decay 37% from its initial intensity after the chair begins to rotate or after it is stopped abruptly, known as the time constant, is recorded.

Static and dynamic SVV require the patient to move a laser image of a line from its current position, which is either left or right of a vertical position, to a vertical position. Static SVV occurs while the patient is sitting still while dynamic SVV occurs while the patient is rotated at a constant velocity at center, slightly to the right, and slightly to the left. While the patient is moved to the left or to the right in the chair, one utricle remains in the center of the axis of rotation while the other utricle is moved “off axis”, which results in this utricle being acted on by horizontal centrifugal forces. This movement is also known as unilateral centrifugation. The off-axis rotation that occurs during unilateral centrifugation activates each utricle in turn. During the off-axis rotations, the patient will experience the sensation of being tilted due to forces being exerted onto the utricles (Brey et al., 2008).

During both static and dynamic SVV, the patient is asked to use two buttons on the chair to orient a line from its current position to what they perceive to be vertical. During dynamic SVV, the patient perceives that he/she is tilting to one direction or the other due to the off-axis rotation. This perception of tilting causes a patient with a normally functioning vestibular system to set the line to the left or right of true vertical (Brey et al., 2008).

II. Anatomy Involved

The rotational chair gives the audiologist insight into the health of the utricles and horizontal semicircular canals; however findings are non-localizing. The superior portions of the vestibular nerves are also involved because the utricles and horizontal SCCs are innervated by this branch. The VOR and the central nervous system's ability to integrate information between the vestibular and visual systems is also involved (Brey et al., 2008)

III. Normal and abnormal findings

SHA is analyzed in regards to gain, phase and symmetry. Patient results are compared with normative data and should fall within a range of normal scores. If phase is normal, the head and eyes are moving the same velocity in opposite directions. If the patient demonstrates phase lead, the eye movement is leading the head movement and if the patient demonstrates phase lag, the eye movement is falling behind the head movement. Gain refers to the average velocity of the slow-phase nystagmus. If high or low gain is noted, it is considered an abnormal finding. Symmetry, which measures the difference between the maximum velocity of the left beat nystagmus or the right beat nystagmus divided by the total of left and right beating slow phase velocity, may be described as normal, leftward, or rightward (Brey et al., 2008).

There are multiple patterns of abnormality that may be noted in SHA. For example, patients who demonstrate phase lead, reduced gain, and abnormal symmetry often have a unilateral, acute peripheral vestibular lesion (Brey et al., 2008) Phase often continues to be abnormal even after the patient compensates for the vestibular lesion, however gain and symmetry will likely return to within normal limits (Baloh & Halmagyi, 1996).

TRAPs are analyzed based on the time constant of the nystagmus and the amount of time, in seconds, that it takes for the nystagmus to decay to 37% of its initial intensity. A normal

finding is for the time constant to be within 10 to 30 seconds. A reduced time constant is considered to be consistent with a peripheral finding (Brey et al., 2008). An increased time constant is considered to be consistent with a central finding or may result from use of medication that alters the central nervous system (CNS) function, such as an anti-depressant, anti-anxiety or sleep medication (Minor et al., 1999).

During static SVV, the patient should orient the line vertically or near vertically for it to be considered a normal finding. During dynamic SVV, when the patient is rotated off-axis to the left, the line should be skewed slightly right to be considered normal. If the patient is off-axis to the right, the line should be skewed slightly to the left to be considered normal. If the patient does not fall within norms when setting the line, the results are considered abnormal. An abnormal result during static and dynamic SVV suggests an uncompensated utricle dysfunction, while a normal finding during static SVV and an abnormal SVV suggests a compensated utricle dysfunction (Brey et al., 2008).

IV. Limitations of the Test and Contraindications to Performing the Test

One limitation of the rotational chair is that ear-specific information cannot be obtained. In addition, if the door to the examination room is not closed completely and light leaks through, the patient will be able to fixate on a visual reference which will lead to high gain during SHA, giving an artifact (Hain, 2014). Patients who are claustrophobic may have difficulty with this evaluation as well because the patient is secured to a chair in a dark room (Brey et al., 2008).

V. Recommendations based on test results

Referrals to otolaryngology may be considered if a peripheral lesion is noted, while referrals to neurology may be considered if central lesions are noted. If the lesion is uncompensated, the patient may be referred for VRT. If rotational chair is being utilized to

monitor for vestibulotoxicity, the audiologist should make the patient's physician aware of any changes in vestibular function (Gillespie & Minor, 1999).

Cervical Vestibular Evoked Myogenic Potential (C-VEMP)

I. Explanation of the Test

The purpose of the Cervical Vestibular Evoked Myogenic Potential (C-VEMP) is to rule out SCD and determine the health of the saccule and the inferior portion of the vestibular nerve (Hain, 2013). The saccule, one of the otolith organs, is sensitive to sound pressure and is stimulated with a high intensity click stimulus and the evoked potential is recorded from the sternocleidomastoid muscle (Akin & Murnane, 2008). One common electrode montage for recording the C-VEMP includes surface electrodes which are placed on the ipsilateral and contralateral sternocleidomastoid muscles (SCM) and on the low forehead. A high-intensity sound stimulus is delivered to the ipsilateral ear with insert earphones. The SCM is contracted for the duration of the evaluation (Colebatch et al., 1994). In order to obtain proper contraction, the patient begins the test in the supine position. The head is turned over the contralateral shoulder and lifted several inches off of the examination table (Gans & Yellin, 2007).

II. Anatomy Involved

The C-VEMP evaluates the VCR and the reflex arc involved is the saccule, the inferior portion of the vestibular nerve, the lateral vestibular nucleus, and to the lateral vestibulospinal tract (Gans & Yellin, 2007).

III. Normal and abnormal findings

The clinician should aim to obtain two replicable and repeatable responses to ensure that the response is valid. The resulting waveform from the C-VEMP is recorded from the SCM and

is a biphasic wave with an initial positive peak known as the P1, followed by a negative peak known as the N1. The P1 is also called the P13 and the N1 is also called the N23, based on the expected latencies. The P1 may be a negative peak and the N1 may be a positive peak, depending on the electrode montage. A normal latency for P1 is between 13 and 20 ms while a normal latency for N1 is located between 20-28 ms. The responses between the ears should be symmetrical, with an asymmetry ratio of less than 33% (Hain, 2013).

As mentioned previously, the saccule is sensitive to sound pressure. A normal threshold response with a click stimulus for the C-VEMP is between 80-100 dB nHL.

It is abnormal to elicit a response to a lower intensity stimulus, often considered to be 70-80 dB nHL. This is considered to be below the normal threshold and is indicative of SCD (Akin & Murnane, 2008). If a C-VEMP is absent, it is important to rule out conductive hearing loss (Hain, 2013). If the C-VEMP is asymmetrical, the patient may have SCD or a unilateral vestibular nerve or saccule disorder.

IV. Limitations of the Test and Contraindications to Performing the Test

Proper patient position is essential in order to obtain accurate test results. The patient is required to maintain a head position that causes contraction of the SCM, which may lead to fatigue especially in older patients. It is important to allow the patient to rest in between each run to ensure valid responses. If a patient has severe neck or back problems, modifications of the patient's position may be required during testing. For example, a pillow may be used to help support the neck or back, or a patient may begin in a semi-reclined position as opposed to supine position.

A C-VEMP may be obtained from a patient with a profound sensorineural hearing loss because the response that is being recorded originates from the saccule, not from the cochlea

(Colebatch et al., 1994). However, a C-VEMP often cannot be recorded if a patient has a conductive hearing loss due to a reduction of sound pressure. The sound pressure is reduced due to the conductive component, such as fluid in the middle or dislocated ossicles, resulting in a reduction of sound pressure reaching the inner ear (Wang & Lee, 2007).

V. Recommendations based on test results

A referral to otology is necessary if SCD is suspected. The otologist may refer the patient for a computed tomography (CT) scan to determine if a dehiscence is present, and if so, the dehiscence may be managed surgically (Telian & Wiet, 2008). The audiologist must also refer the patient to otology if an asymmetry in amplitudes is noted. The C-VEMP is either absent, or the amplitude of the waveform is decreased, in approximately 80% of patients with a vestibular schwannoma. Similarly, an absent C-VEMP or reduced amplitude is noted in up to 72% of patients with vestibular neuronitis (Akin & Murnane, 2008).

Posturography

I. Explanation of the Test

Posturography is used to evaluate postural stability when one or more of the three balance systems (vestibular, visual, and somatosensory) are compromised (Nashner, 1997). During posturography, the patient is secured to a harness and stands on a platform, facing a visual surround. Arms are at the patient's side and he/she is instructed to stand upright and maintain balance throughout the evaluation. The platform is sensitive to the patient's weight distribution, therefore sensitive to any movement or changes in the patient's center of gravity (Neurocom, 2012).

The platform and visual surround are “sway-referenced”, meaning that they will move or tilt with the patient’s body movement. This removes the ability for the patient to rely on either the somatosensory or visual systems, causing them to rely on the other systems that are not compromised in order to maintain balance. The clinician is able to analyze whether the patient makes appropriate reflexive compensatory movements to recover balance during this evaluation. There are six different conditions, increasing in complexity, which compromise one or more of the three balance systems (Neurocom, 2012). See Figure 1 for a list of the six conditions.

II. Anatomy Involved

Posturography evaluates postural stability in regards to the ability of the vestibular, somatosensory, and visual systems to work together and how the patient is able to maintain balance when one or more of these systems is compromised (Nashner, 1997).

III. Normal and abnormal findings

The normative data in posturography evaluation is based on age. It is considered normal for the patient to maintain balance while demonstrating a minimal amount of sway throughout each of the six conditions. Excessive sway or falls are considered abnormal (di Fabio, 1995).

Specific patterns of abnormalities within different conditions have been found to relate to deficits within one of the three balance systems. Patients who demonstrate abnormalities in conditions 5 and 6 are said to have a vestibular deficit. Individuals who demonstrate difficulties with conditions 2, 3, 5 and 6 depend highly on their visual systems while patients who demonstrate abnormal results in conditions 4, 5 and 6 have a dependence on the somatosensory system for balance. Individuals who demonstrate difficulties on conditions 1 and 2 while performing normally on subsequent conditions may be suspected of malingering (di Fabio, 1995).

Patients with peripheral vestibular lesions may demonstrate abnormal sway or falls when both the somatosensory and visual systems are compromised simultaneously. In the acute phase of the lesion, the patient may demonstrate abnormal test results when they are forced to rely only on the vestibular system to maintain balance. If the patient has centrally compensated to the vestibular lesion, they may demonstrate abnormalities in other vestibular evaluations such as bithermal calorics or rotational chair, however he/she may demonstrate normal results during posturography.

IV. Limitations of the Test and Contraindications to Performing the Test

One limitation of posturography is that the results will not determine a specific site of lesion; rather the test is used to evaluate the patient's ability to maintain balance while under a variety of different conditions (Furman, 1994). The clinician may not be able to perform this test if the patient has weakness or limited mobility of the ankles or hips (Nashner, 1997), or if the patient is unable to stand without the aid of a cane or a walker for a prolonged period of time. The patient is required to stand and maintain balance throughout a variety of different conditions during this evaluation. Weakness of the hips or ankles as well as the inability to stand unaided for a prolonged period of time may impede the patient's ability to complete this evaluation.

IV. Recommendations based on test results

Patients who demonstrate abnormalities during posturography may be referred for VRT to help achieve compensation (Boyer et al., 2008). The clinician may also discuss fall-risk management strategies with the patient, such as eliminating clutter in the house, using a seat in the shower, or ensuring that the house is well-lit.

CHAPTER FOUR

CASE STUDIES

Case Number One:

I. Case History

B.D., a 74 year old female, was referred from otology to audiology for a vestibular evaluation including VNG and rotational chair. She reported that she experienced an episode of true vertigo, lasting minutes, which woke her from sleep approximately six years ago. Ms. D was reportedly given meclizine to treat the vertigo, however she denied subjective improvement. Since that time, she has experienced what she described as constant imbalance and feeling as though she veers to the left when walking. B.D. stated that she no longer drives due to her symptoms. She denied nausea, vomiting, and falls associated with her imbalance. She also denied head trauma or illness immediately prior to the episode of vertigo six years ago.

Ms. D. also reported vertigo, lasting seconds, when she lays on the right side of her body or puts her head back, like when she is having her hair washed at a beauty salon. She reportedly avoids the provoking positions. She also reported aural fullness on the right side. B.D. denied tinnitus.

Ms. D denied a history of stroke, seizures and migraines. She reported that she is pre-diabetic and has atrial fibrillation. B.D. denied the intake of caffeine, alcohol and medications 48-hours prior to the evaluation.

II. Test Results

Results are displayed beginning on page 68. Tympanometry revealed normal ear canal volume, compliance, and middle ear pressure bilaterally (Fig. 2A). This is suggestive of a normal middle ear system for bithermal caloric evaluation, bilaterally.

SHA revealed reduced gain, phase lead, and a rightward asymmetry (Fig 2B). TRAPs revealed normal time constants (Fig 2C). A leftward preference was noted during static SVV (Fig 2D). Dynamic SVV was not performed due to nausea and discomfort reportedly resulting from the vestibular assessment and she requested that the testing be discontinued.

No spontaneous nystagmus was noted during the VNG evaluation. Saccades, OPKs, smooth pursuit (Fig 2E) and gaze testing results (Fig 2F) were unremarkable. The high-frequency headshake was unremarkable (Fig 2G). The Dix-Hallpike demonstrated geotropic rotary nystagmus in the head right condition and nystagmus reversed direction upon returning to the seated position (Fig 2G). This is suggestive of right posterior canal BPPV.

Positional evaluations were unremarkable, however patient reported transient vertigo (Fig 2H). Bithermal caloric evaluation demonstrated a 30% weakness of the right ear (Fig 2I).

III. Interpretation and Recommendations

Static SVV suggested an uncompensated utricle dysfunction. Previous studies have suggested that patients with BPPV may also have utricle dysfunction because degeneration of the utricle may cause the otoconia to come loose from the otolithic membrane (Seo et al., 2013). The unilateral weakness noted during caloric evaluation was suggestive of a right-sided peripheral vestibulopathy. Geotropic nystagmus noted during the Dix-Hallpike is suggestive of right posterior canal BPPV. It was recommended that Ms. D follow-up with otology, undergo the Epley maneuver due to posterior canal BPPV, and consider VRT if symptoms persist to aid with central compensation of the unilateral weakness.

Case Number Two:***I. Case History***

D.D., a 57 year old female, was referred from otology to audiology for a vestibular evaluation including VNG. Otologic history was significant for Meniere's disease of the right ear, which was diagnosed in 2009. D.D. denied a family history of Meniere's disease. In 2011, she underwent an endolymphatic sac decompression of the right ear with subjective improvement following surgery. She also reported that she was diagnosed with BPPV in the right ear immediately following surgery and for approximately one year after that. She reportedly had multiple Epley maneuvers performed on her by a vestibular therapist and performed several maneuvers at home on her own. The BPPV had since resolved. D.D. had a VNG evaluation completed approximately two years prior to the current testing. Caloric results demonstrated a 37% unilateral weakness to the right, consistent with the previous diagnosis of Meniere's disease.

Approximately one month prior to the current evaluation, Ms. D. reportedly began to experience Meniere's attacks again. She described her attacks as vertigo accompanied by nausea and emesis that lasted for hours, aural pressure in the right ear only, and hearing fluctuations in the right ear only. She took meclizine as needed during these attacks, sometimes as often as five times per day. Ms. D. received intra-tympanic and oral steroids with subjective improvement reported and her attacks were less frequent and less severe since beginning this course of treatment.

D.D. reported a constant, pulsating tinnitus in the right ear only, which began following her endolymphatic sac decompression surgery in 2011. Ms. D was found to have asymmetric hearing

loss, with the hearing in her right being poorer than left, however she reported that she has never worn a hearing aid.

Ms. D. denied a history of diabetes, stroke, seizures and migraines. She stated that she was diagnosed with high blood pressure approximately 15 years ago; however it is controlled with medication. D.D. denied the intake of alcohol, caffeine and medications 48-hours prior to the current evaluation.

II. Test Results

Tympanometry revealed normal ear canal volume, compliance, and middle ear pressure bilaterally. This is suggestive of a normal middle ear system for bithermal caloric evaluation, bilaterally (Fig 3A). The audiometric evaluation revealed an asymmetric hearing loss, right poorer than left, consistent with history of Meniere's disease (Fig 3B).

No spontaneous nystagmus was noted during the VNG evaluation. Saccades and OPK testing were unremarkable. Catch-up saccades were noted during smooth pursuit (Fig 3C). LBN (approximately 2deg/sec) was noted in the gaze left condition. Nystagmus suppressed with fixation (Fig 3D). LBN (approximately 2deg/sec) was noted after the high-frequency headshake (Fig 3E). The Dix-Hallpike was unremarkable and subjective vertigo was denied (Fig 3E). Slight LBN (approximately 3deg/sec) was noted in the head left position. Nystagmus suppressed with fixation. All other positions were unremarkable (Fig 3F). Bithermal calorics revealed a 50% unilateral weakness to the right with reduced overall caloric responses on the right and normal caloric responses on the left (Fig 3G).

III. Interpretation and Recommendations

The purpose of this evaluation was to determine if the patient's Meniere's disease has become a bilateral issue and was now impacting her left ear as well because she had gone several

months without experiencing a Meniere's attack. The combined results are suggestive of a right peripheral vestibulopathy, consistent with diagnosis of Meniere's disease on the right and previous vestibular evaluation which revealed a right-sided vestibular weakness. LBN noted throughout testing suggests that the vestibulopathy is in the active, non-compensated state. D.D. was referred to otology and vestibular rehabilitation therapy was recommended.

This test was important because, as mentioned previously, Meniere's disease is bilateral in approximately 24% of patients and the second ear presents an average of 7.6 years following the first ear (House et al., 2006). It is a positive indicator in this patient that the second ear does not seem to be impacted at this time. She was referred to vestibular rehabilitation to help central compensation for a unilateral peripheral vestibular weakness.

Case Number Three:

I. Case History

S.M., a 28 year old female, was referred from otology to audiology for a vestibular evaluation including rotational chair and VNG. She was diagnosed with bilateral hearing loss at birth. The hearing loss was progressive and she wore hearing aids until age 11 years, when she post-lingually received a cochlear implant (CI) in the left ear. Approximately ten years later, the patient received a CI in the right ear as well. She reported that she was dizzy for approximately ten days following implantation of the right ear; however the dizziness subsided after the ten days. Ms. M. denies any current imbalance or dizziness.

Ms. M has been implanted in the left ear for approximately 20 years and the right ear for approximately ten years. She was examined by her otologist due to recent onset of pain at the implant site in the right ear. Ms. M. was evaluated by the audiologist who follows her for her CI

and it was determined that the magnet strength was appropriate. S.M. underwent a vestibular evaluation because she wishes to have the right side explanted and re-implanted to relieve pain at the implant site. She was referred to have a vestibular evaluation to determine vestibular function prior to re-implantation.

Ms. M. denied a history of high blood pressure, diabetes, stroke, seizures, and migraines. She denied the intake of caffeine, alcohol and medications 48 hours prior to her vestibular evaluation.

II. Test Results

Tympanometry revealed normal ear canal volume, compliance, and middle ear pressure bilaterally. This is suggestive of a normal middle ear system for bithermal caloric evaluation, bilaterally (Fig 4A).

SHA revealed reduced gain and phase lead overall (Fig 4B). TRAPs revealed reduced time constants (Fig 4C). Combined results from SHA and TRAPs are suggestive of a peripheral vestibulopathy. Static and dynamic SVV demonstrate a rightward preference, suggestive of uncompensated utricle involvement (Fig 4D).

Spontaneous LBN, approximately two degrees per second, was visualized throughout VNG evaluation. All nystagmus suppressed with fixation. Oculomotor and gaze testing were unremarkable (Fig 4E and 4F). Minimal RBN was noted immediately post-headshake and following RBN fatigue, spontaneous LBN was noted (Fig 4G). The Dix-Hallpike was unremarkable bilaterally (Fig 4G). Spontaneous LBN was noted in most positions, however all nystagmus suppressed with fixation (Fig 4H).

Bithermal caloric evaluation revealed a 100% unilateral weakness to the left. No response greater than spontaneous nystagmus was noted from bithermal calorics on the left side. Ice water calorics were performed on the left side to ensure no response. No nystagmus greater than

spontaneous nystagmus was noted post ice-water calorics. This was suggestive of left sided canal paralysis. Responses for the right side were reduced for patient age (Fig 4I).

III. Interpretation and Recommendations

Combined results were suggestive of bilateral peripheral vestibulopathy including otolith involvement, with the left ear significantly poorer than the right ear. Bithermal caloric evaluation demonstrated canal paralysis of the left ear. The right ear, which was the ear that the patient is considering cochlear re-implantation, was the only ear with residual vestibular function. If vestibular function were to be damaged during surgery, the patient would risk having no vestibular function bilaterally. It was recommended that patient return to otology with vestibular test results to discuss re-implantation risks and benefits based on her vestibular function. Acute peripheral vestibular dysfunction occurs in 30-60% of patients as a result of the trauma caused by cochlear implantation, and chronic dizziness occurs in approximately one-third of cochlear implant recipients (Fina et al., 2003). Patients with bilateral peripheral dysfunction experience constant imbalance, unsteadiness, and oscillopsia while moving due to disruptions in the VOR (Funabashi et al., 2012).

Case Number Four:

I. Case History

S.R., a 61 year old male, was referred from otology to audiology for a vestibular evaluation including C-VEMP, rotational chair, and VNG. Mr. R. was diagnosed with an acoustic neuroma on the right side one month prior to his vestibular evaluation. The purpose of the vestibular evaluation was to determine the residual function of the vestibular portion of CNVIII in order to

aid in the determination of the surgical approach- nerve preservation versus sacrifice of the nerve.

Mr. R. reported feelings of imbalance and clumsiness beginning six months prior to his vestibular evaluation and are gradually worsening. He also stated that he tends to sway when walking and will collide with walls and doorways. He denied nausea, emesis, vertigo, and falls related to his symptoms.

S.R. also reported sudden hearing loss of the right ear, occurring approximately three months prior to his vestibular evaluation. He stated that sounds in that ear have a “mechanical” or “cartoonish” quality to them. An audiogram at an outside facility revealed that Mr. R. has 100% word recognition in the left ear and 52% word recognition in the right ear. He also reported tinnitus which he described as a “hissing” noise in the right ear only. Mr. R. denied a history of high blood pressure, diabetes, seizures, migraines, or stroke. He also denied the intake of caffeine, alcohol and medications 48 hours prior to his vestibular evaluation.

II. Test Results

Tympanometry revealed normal ear canal volume, compliance, and middle ear pressure bilaterally. This is suggestive of a normal middle ear system for C-VEMP and bithermal caloric evaluation (Fig 5A).

C-VEMP testing revealed an absent response from the right side, suggesting abnormal function of the right saccule or inferior branch of the vestibular nerve. The asymmetry ratio is 100%. This is consistent with patient’s diagnosis of right acoustic neuroma (Fig 5B).

SHA testing revealed reduced gain and phase lead, which is a peripheral finding and is consistent with the diagnosis of a right acoustic neuroma (Fig 5C). TRAPs revealed essentially

reduced time constants, which is also a peripheral finding and consistent with the patient's diagnosis (Fig 5D).

Spontaneous RBN, approximately 4 degrees per second, was noted during center gaze and suppressed with fixation with VNG testing. Saccades, smooth pursuit, and OPKs were unremarkable. RBN, approximately two degrees per second, was noted in gaze right. Nystagmus was noted in all other gaze positions as well; however nystagmus was no greater than spontaneous and suppressed with fixation. These results are consistent with an uncompensated right peripheral vestibulopathy and with patient diagnosis of right acoustic neuroma. See Figures 5e and 5f for oculomotor and gaze testing.

Slight LBN, too minimal to mark, was noted immediately post-headshake. Dix-Hallpike was unremarkable bilaterally and patient denied subjective vertigo. These results are consistent with patient diagnosis (Fig 5G).

Minimal geotropic nystagmus was noted in all positions during positional evaluations and nystagmus suppressed with fixation in all positions. This is consistent with the patient's diagnosis (Fig 5H).

Bithermal caloric responses revealed a 77% caloric weakness to the right, which is consistent with patient diagnosis of a right acoustic neuroma (Fig 5I).

III. Interpretation and Recommendations

Combined responses are suggestive of a right peripheral vestibulopathy, which is consistent with the diagnosis of a right acoustic neuroma. The responses from the right ear are significantly reduced compared to the responses from the left ear. It was recommended that S.R. follow-up with otology regarding medical management of the acoustic neuroma.

The otologist made the decision to perform surgery with a cranial fossa approach rather than a translabyrinthine approach due to the results from the audiogram and the vestibular evaluation. The translabyrinthine surgical approach always involves sacrifice of CN VIII, while the cranial fossa approach has up to a 71% chance of preserving the nerve, based on the size and location of the tumor (Baumann et al., 2005). Both the audiogram and the vestibular evaluation revealed that Mr. R. still has function in his right CN VIII so the otologist made the decision to try to preserve, rather than sacrifice, the nerve during surgery.

Case Number Five:

I. Case History

P.P., a 50 year old female, was referred from otology to audiology for a vestibular evaluation including a VNG. She reported that she was in good health prior to an episode where she had the flu approximately four months prior to the vestibular evaluation. Two weeks following the illness, she reportedly began to experience significant imbalance and disequilibrium. She experienced true vertigo, lasting seconds, which occurred when laying down or getting up. P.P. has not had episodes of true vertigo since the month following her illness. Several weeks after the flu, Ms. P. reported that she began to experience a tremor, lack of coordination, slurred speech and lack of ability to speak, and nausea as well as emesis.

She was admitted to the hospital at that time and again several weeks later. She began to ambulate with a wheelchair at that time and was unable to walk or stand without assistance. P.P. was evaluated by a neurologist, who stated that her symptoms are due to an infection of the

cerebellum, which was determined via lumbar puncture. CT scans and MRI scans were reportedly unremarkable.

Ms. P. had recovered the ability to speak since the onset of her symptoms. Her tremors decreased in intensity. She denied diabetes, history of seizures, and history of stroke, light and sound sensitivity, hearing loss, aural fullness, previous otologic surgeries, head trauma, and migraines. She reported a positive family history of Meniere's disease. P.P. denied the intake of caffeine and alcohol 48-hours prior to the vestibular evaluation. She regularly takes medication for her tremors, however discontinued three days prior to this evaluation.

II. Test Results

Tympanometry revealed normal ear canal volume, compliance, and middle ear pressure bilaterally. This is suggestive of a normal middle ear system for bithermal caloric evaluation, bilaterally (Fig 6A).

No spontaneous nystagmus was noted during the VNG evaluation. Eye movement was noted throughout testing secondary to involuntary tremors. Increased latencies and reduced accuracy were noted during saccades. Patient demonstrated saccadic catch-ups throughout smooth pursuit testing. Decreased gain was noted during OPK testing (Fig 5B).

Gaze testing (Fig 5C), high-frequency head shake (Fig 5D), Dix-Hallpike (Fig 5D), and positional testing (Fig 5E) were unremarkable. Bithermal caloric results demonstrated hyperactive vestibular responses bilaterally and a high fixation index (Fig 5F).

III. Interpretation and Recommendations

The purpose of the vestibular evaluation was to determine if the patient's symptoms were due to the cerebellar infection or if she had a comorbid peripheral vestibular dysfunction. This was of significant importance due to a positive family history of Meniere's disease. Abnormal

oculomotor testing, bilateral hyperactive caloric responses, and a high fixation index were suggestive of central involvement. No peripheral indicators were noted at the time of the vestibular evaluation.

These results are significant because it is important to consider possible comorbidities, especially in difficult cases such as this. Comorbidities may be overlooked if the health care providers are only focused on one known cause of dizziness. As mentioned previously in this document, approximately 10% of patients with Meniere's disease have a family history of Meniere's disease (Fung et al., 2002). With a positive family history of Meniere's disease, plus a history of vertigo as a symptom, it is important to rule out peripheral causes of dizziness in this patient. This patient has a known central cause of dizziness, however if a peripheral balance disorder was discovered and treated it may have improved this patient's quality of life. Since no peripheral disorder was found, it was recommended that this patient follow-up with her neurologist.

CHAPTER FIVE

CONCLUSION

The vestibular system is complex in terms of anatomy and physiology and many audiologists do not understand the vestibular system as completely as they understand the auditory system. It is important for audiologists to understand the anatomy and physiology of the vestibular system as well as have a basic understanding of the tools that are available to audiologists to aid in the differential diagnosis of vestibular disorders.

As stated previously in this document, it is within the audiologist's scope of practice to aid in the identification and management of vestibular disorders. Audiologists are one of the few health care providers who are uniquely trained in the anatomy and physiology of the vestibular system as well as the symptomology of different vestibular disorders.

It is unrealistic to expect every audiology clinic to have the equipment required to perform a full vestibular test battery as these machines are very expensive and take up a significant amount of space. However, all audiologists should have some basic knowledge of the vestibular system and some common disorders in order to help point dizzy patients in the right direction for appropriate care.

All patients should be asked if they experience dizziness as a symptom during their case history, whether or not the audiologist that examines them has the equipment required for a vestibular evaluation at their facility. Patients may not offer this information if they are not directly asked, especially if they are being evaluated for hearing disorders. The patient may be unaware that the ear is related to balance disorders as well. If the patient answers positively to this question stating that they do experience dizziness, the audiologist should continue with a brief case history regarding the dizziness.

There are several questions that should be asked of the patient in order to differentiate possible disorders. The patient should first be asked to describe their symptoms without using the term “dizzy”. As stated earlier in this document, it is important to determine if the patient has experienced vertigo in order to distinguish between vestibular disorders and non-vestibular dizziness. The patient should also be asked how many episodes of dizziness they have encountered and the duration of the episodes. These questions will help distinguish between certain disorders as well, because different disorders present with different symptoms. The patient should be asked to recall any triggers that provoke the episodes; such as movement, pressure changes, or loud sounds. Any possible medical or pharmacological influences should also be explored. This basic six-question case history will help guide the audiologist to which professional the patient is referred.

Certain tests may be performed by audiologists with standard audiology diagnostic equipment on patients who are complaining of dizziness. Dizzy patients should be asked if tympanometry provokes their symptoms. If the patient states that it does, the audiologist should note this in their records as they may have a perilymphatic fistula. If the patient states that loud noises provoke their symptoms, the audiologist may present high-intensity sounds through the audiometer and ask the patient to report any dizziness. A positive result may be indicative of SCD. Audiologists should also perform bone conduction testing at all frequencies including 250 Hz for patients who are complaining of dizziness because SCD may result in a low-frequency conductive hearing loss, as stated earlier in this document.

In addition, audiologists must be aware of certain patterns that may present in an audiometric evaluation. For example, if a patient presents with vertigo, tinnitus, aural fullness and hearing loss, and the audiogram reveals a low-frequency hearing loss, the audiologist must

refer the patient to an otologist to rule out Meniere's disease. If a patient presents with asymmetric high-frequency hearing loss and complains of general imbalance, the audiologist must refer the patient to an otologist to rule out retrocochlear pathology such as an acoustic neuroma. An audiologist does not need to be a vestibular specialist in order to recognize patterns in symptomology and audiometric test results and make appropriate referrals.

For audiologists who do perform vestibular evaluations, it is crucial to have an in-depth knowledge of vestibular disorders, symptomology, what tests to perform, when to add additional tests (ex: Fistula test) outside of the normal test battery, and what the results should look like. For example, an audiologist who only has access to VNG equipment should be aware of the limitations of the VNG and be able to make referrals for additional vestibular testing at an outside clinic when warranted. For example, the audiologist may consider a referral to an outside clinic for rotational chair evaluation if the VNG is normal but a high-frequency peripheral vestibulopathy may be present. Vestibular audiologists should also be comfortable making referrals to other professionals, such as otologists, neurologists, vestibular therapists, or audiologists with different vestibular equipment, when necessary. Management of a dizzy patient will often require a multi-disciplinary approach.

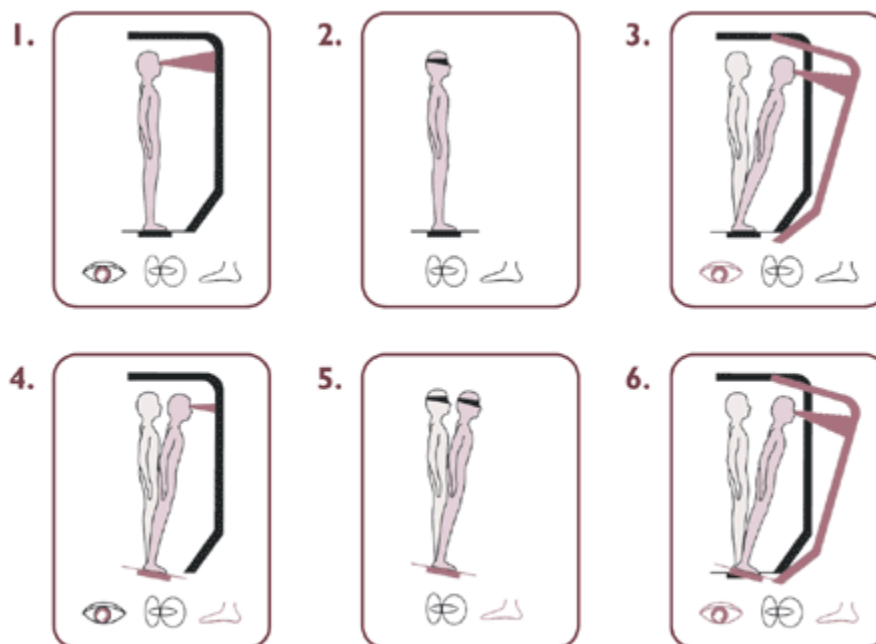
Vestibular disorders may have a large negative impact on a patient's life, causing them to be unable to drive, unable to lie on a certain side in bed, or causing them to feel nauseous or vomit. Some patients may be in danger with vestibular dysfunction, especially an elderly patient with brittle bones who may fall and break a bone because of their balance disorder. Often, patients will be seen by multiple different providers and are put on vestibular suppressants, such as meclizine, long term and are never truly treated for the cause of their dizziness. Audiologists are considered to be experts in the area of balance disorders, and all audiologists have a

responsibility to their patients to provide appropriate care in the area of balance or to provide appropriate referrals. Proper diagnosis and treatment of balance disorders may greatly improve a patient's quality of life.

The purpose of this document was to give an overview of the anatomy and physiology of the vestibular system, discuss some common disorders, provide an overview of the diagnostic tools available to audiologists, and to discuss some interesting vestibular cases. It is essential for all audiologists, even those who do not perform vestibular evaluations, to have a working knowledge of the vestibular system, common vestibular disorders and symptoms reported, and the vestibular evaluations that are used to aid in differential diagnosis. This knowledge will aid audiologists in making appropriate referrals for their patients to obtain proper care.

FIGURES

1. Posturography Conditions

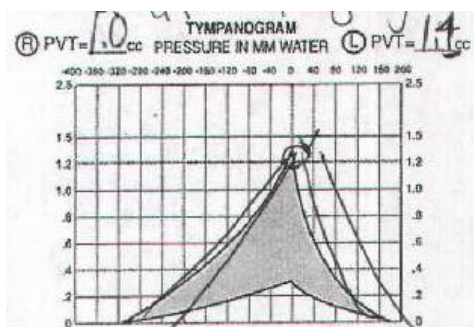


Sensory Organization Test

Description: This image depicts the six conditions that are completed in the sensory organization test in posturography. In the first condition, the platform and visual surround are stable and the patient has their eyes open. In the second condition, the platform and visual surround are stable and the patient has their eyes closed. In the third condition, the platform is stable and the visual surround is sway-referenced. In the fourth condition, the platform is sway-referenced and the visual surround is stable. In the fifth condition, the platform is sway-referenced and the patient has their eyes closed. In the sixth condition, both the platform and the visual surround are sway-referenced. Adapted from Computerized Dynamic Posturography Protocols, by NeuroCom, retrieved from <http://resourcesonbalance.com/program/role/cdp/protocols.aspx>. Copyright 2012 by NeuroCom.

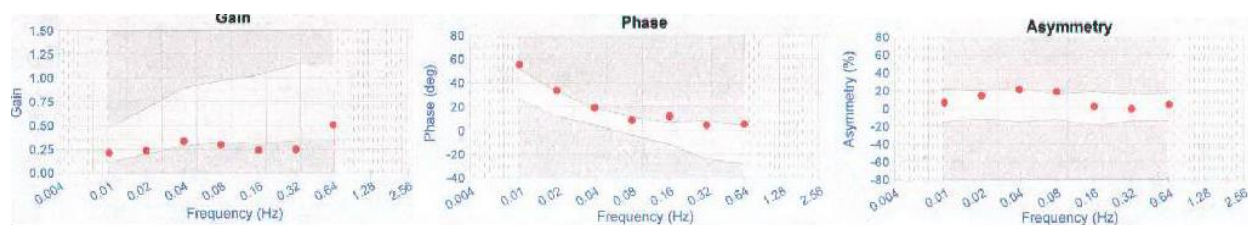
2. Case Number One: B.D.

a. Tympanometry



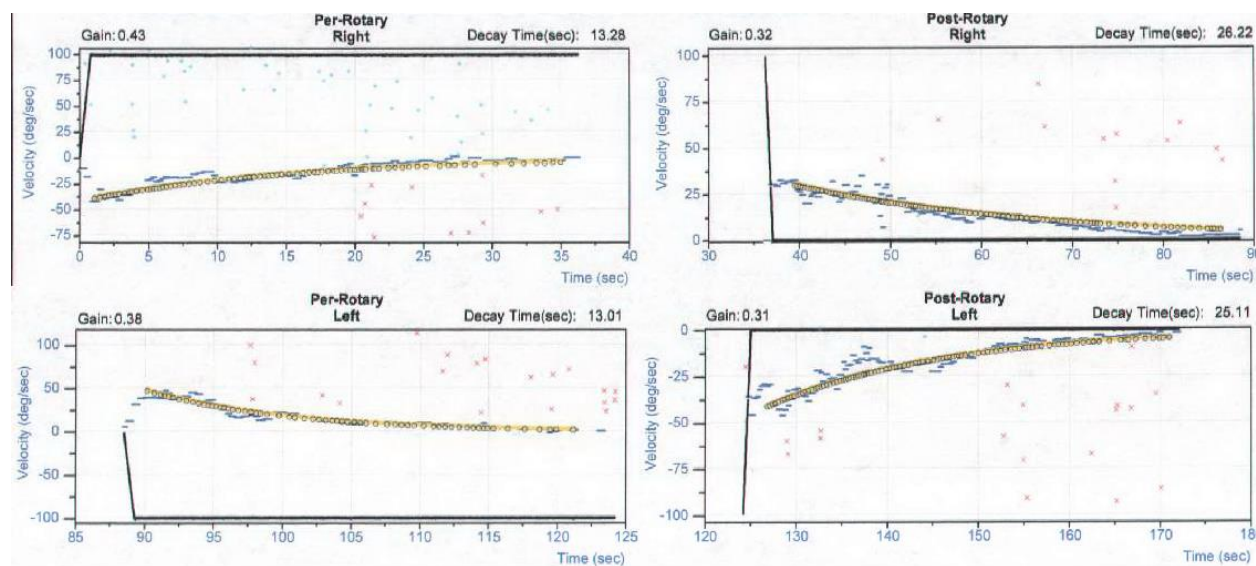
Description: Normal ear canal volume, compliance, and middle ear pressure, bilaterally. This suggests a normal middle ear system for bithermal caloric evaluation, bilaterally.

b. Rotational Chair: SHA



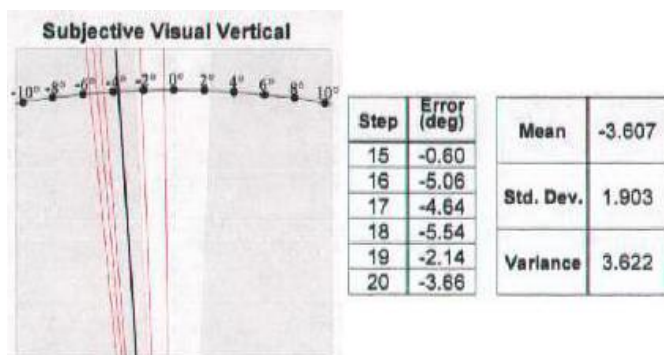
Description: Reduced gain and phase lead with an asymmetry to the right. This is suggestive of a peripheral vestibulopathy.

c. Rotational Chair: TRAPs



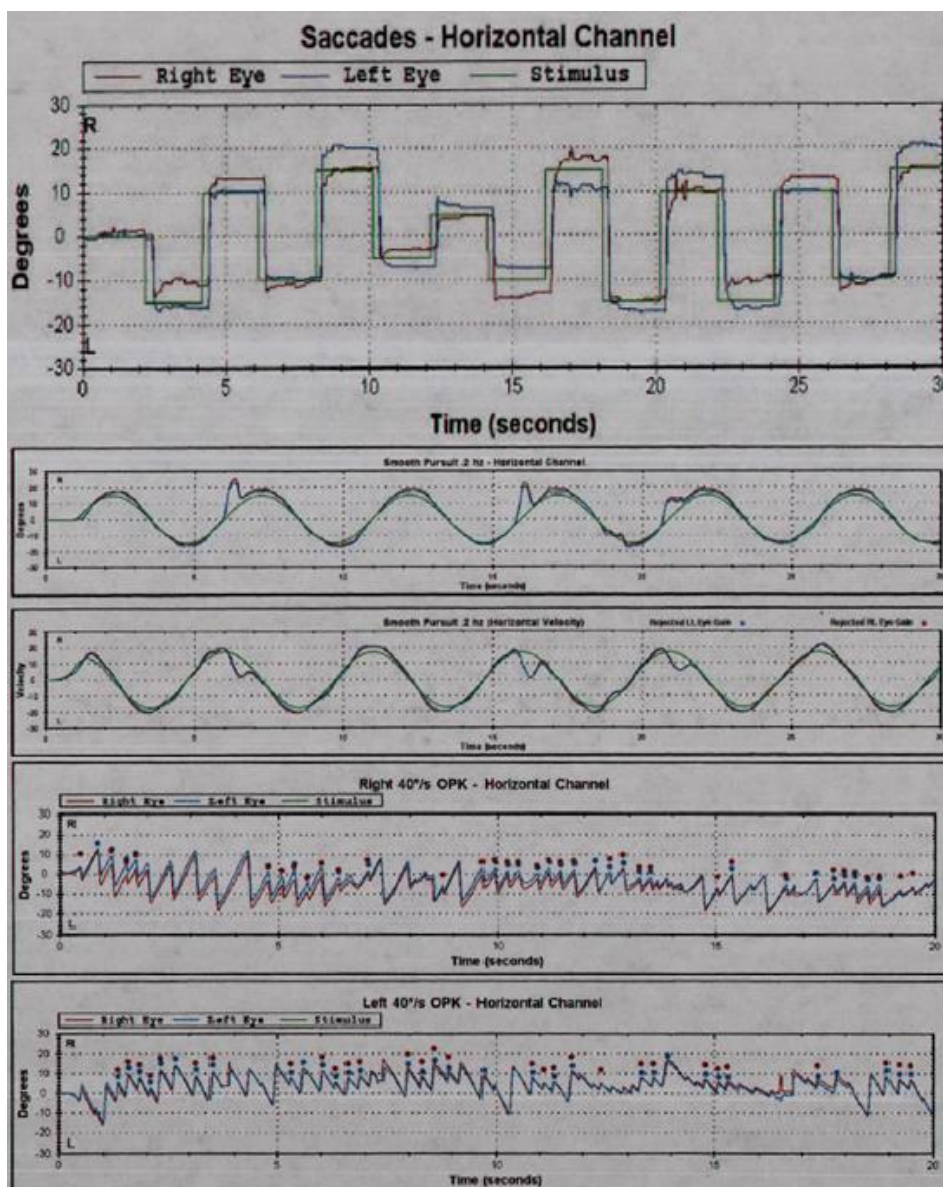
Description: Normal time constants.

d. Rotational Chair: Static SVV



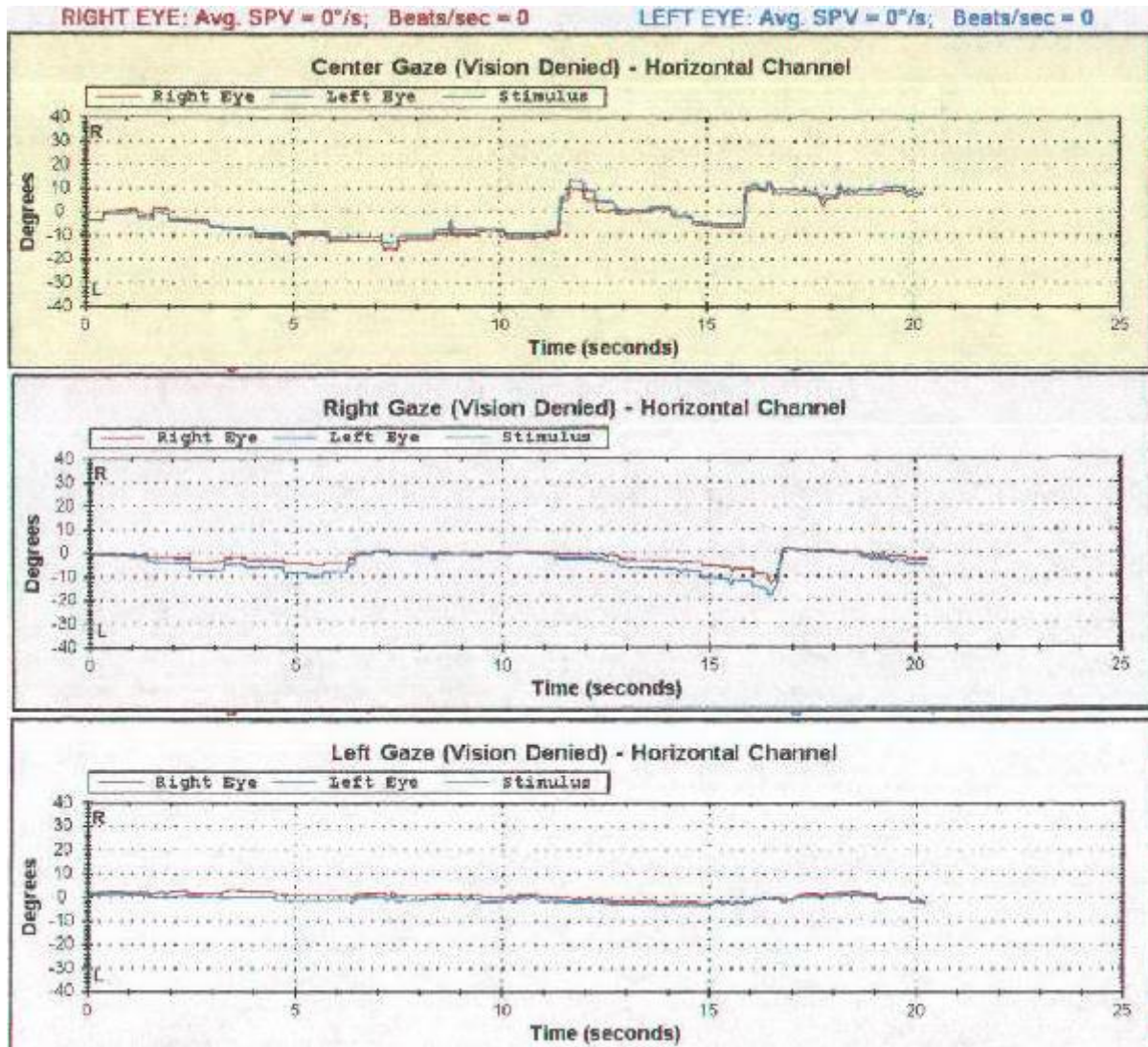
Description: Leftward preference, suggestive of an uncompensated utricle dysfunction.

e. VNG: Oculomotor Evaluation



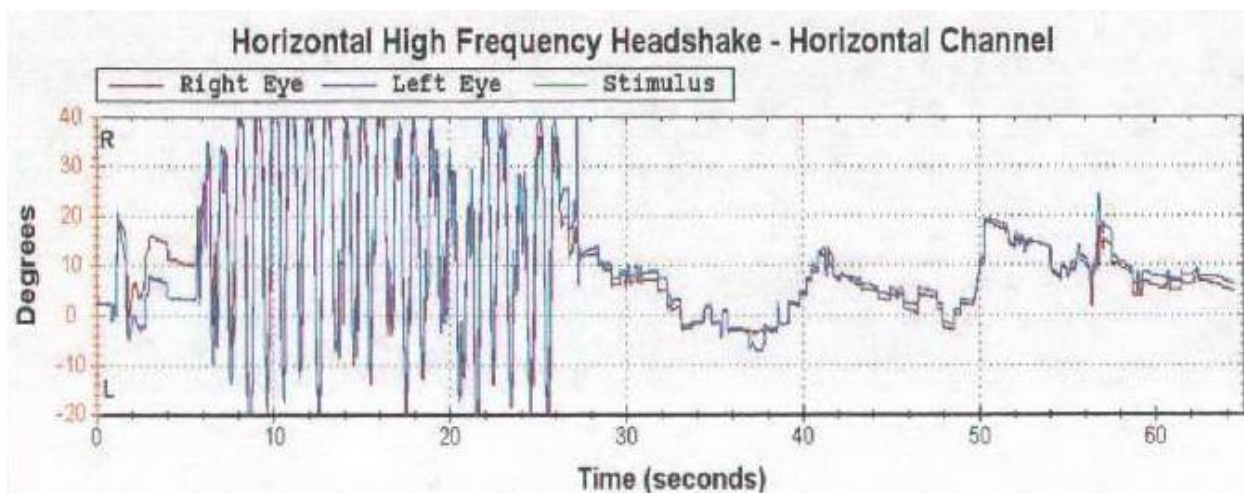
Description: Oculomotor testing was unremarkable.

f. VNG: Gaze testing, vision denied.

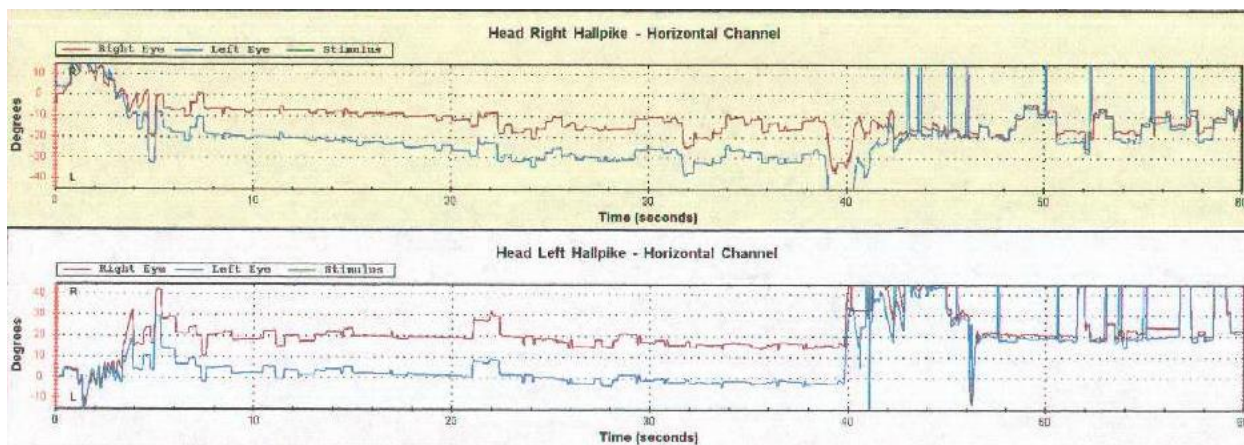


Description: Gaze testing is unremarkable.

g. VNG: High-frequency headshake and Dix-Hallpike

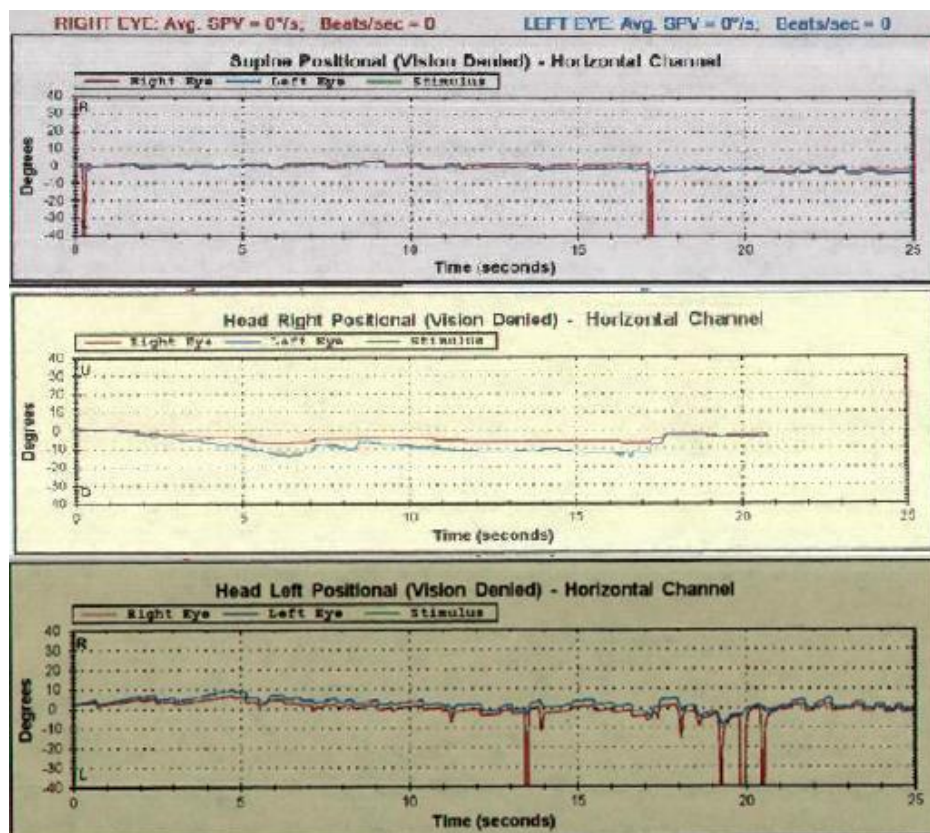


Description: High-frequency headshake is unremarkable.



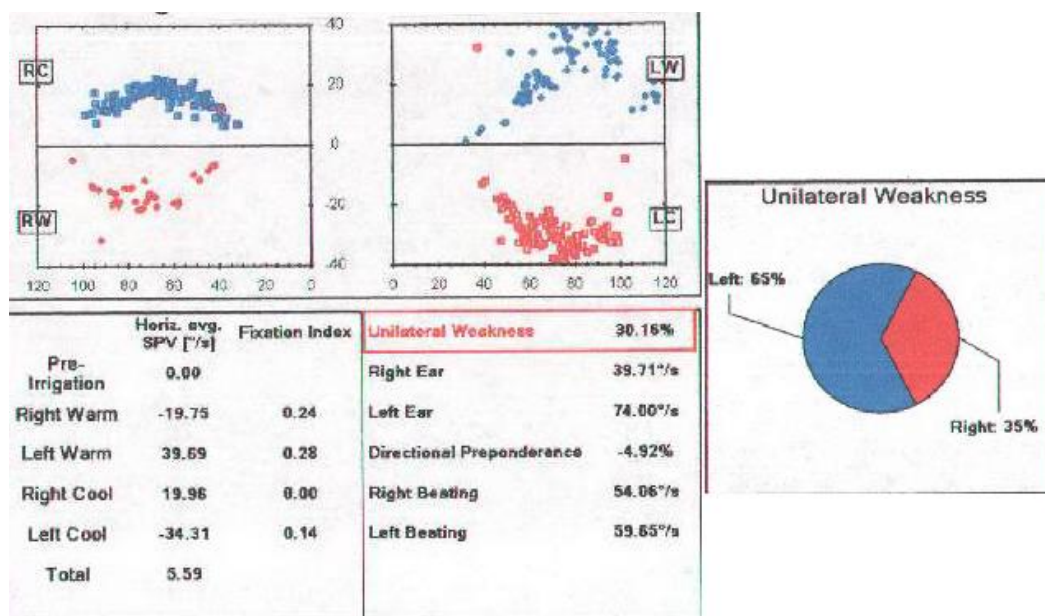
Description: Geotropic, rotary nystagmus was noted during Dix-Hallpike head right. Transient vertigo was reported. Dix-Hallpike head left was unremarkable.

h. VNG: Positional Evaluations



Description:
Positional testing
was unremarkable.

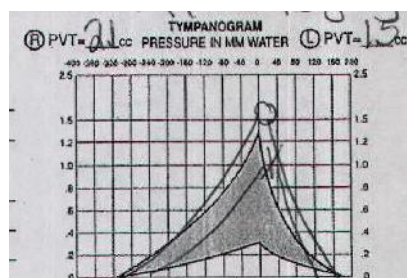
i. VNG: Bithermal caloric evaluation



Description: Bithermal caloric evaluation revealed a 30% right-sided unilateral weakness.

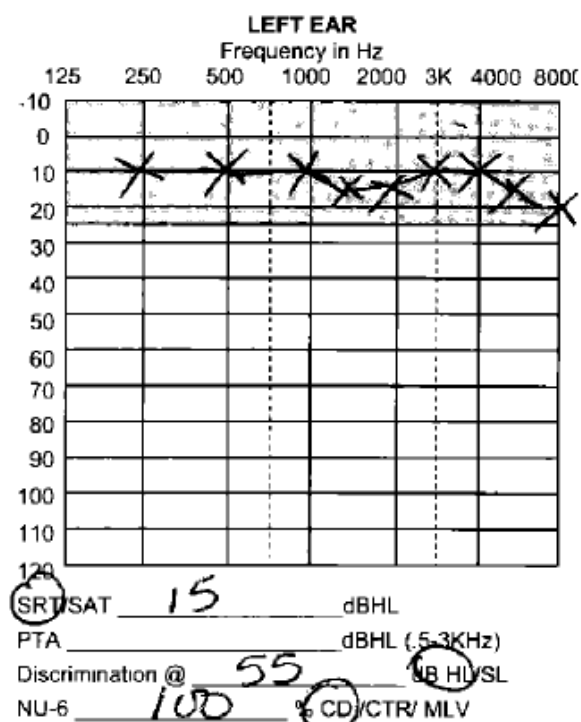
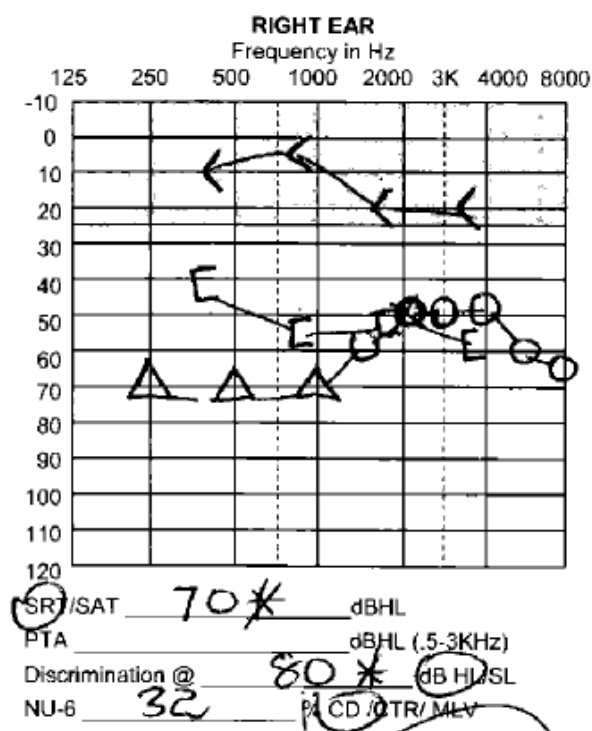
3. Case Number Two: D.D.

a. Tympanometry

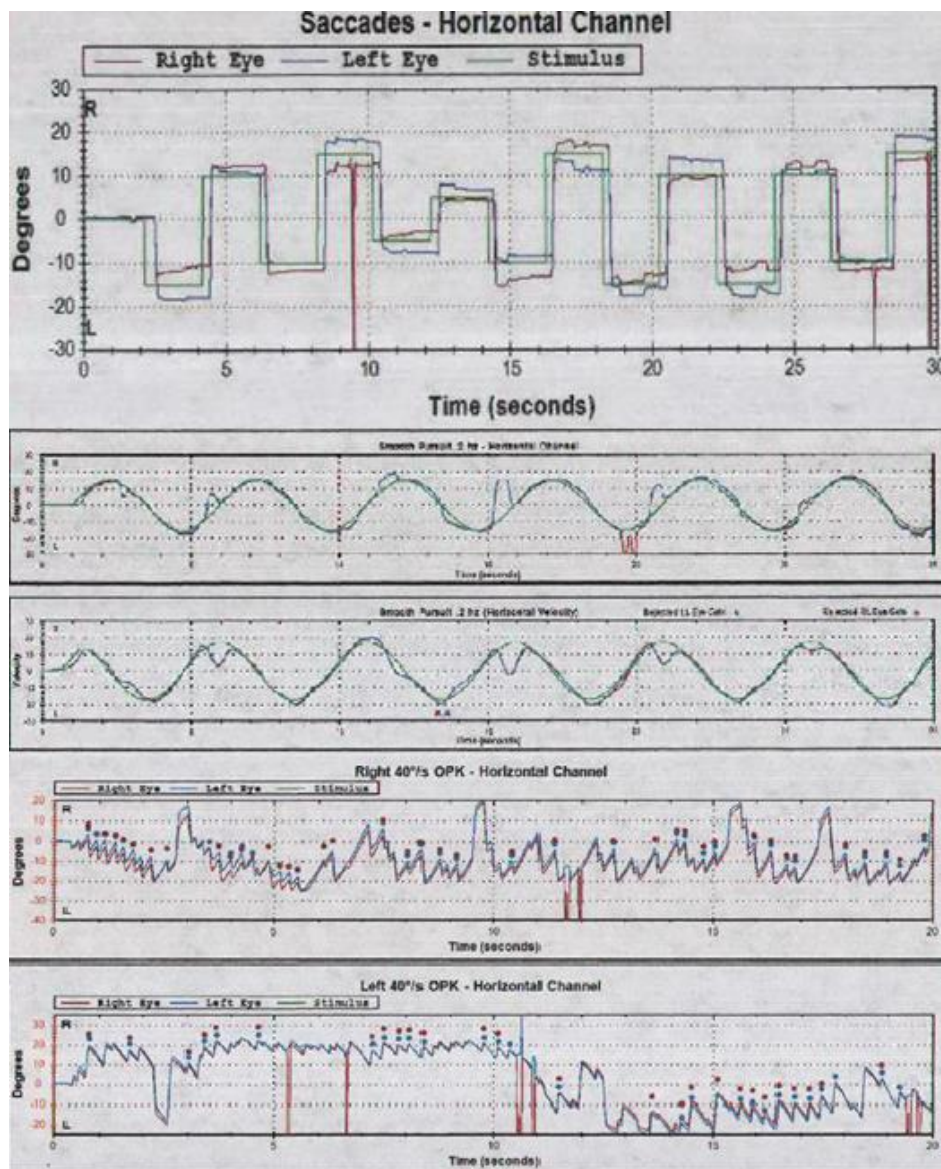


Description: Normal ear canal volume, compliance, and middle ear pressure, bilaterally. This suggests a normal middle ear system for bithermal caloric evaluation, bilaterally.

b. Audiogram

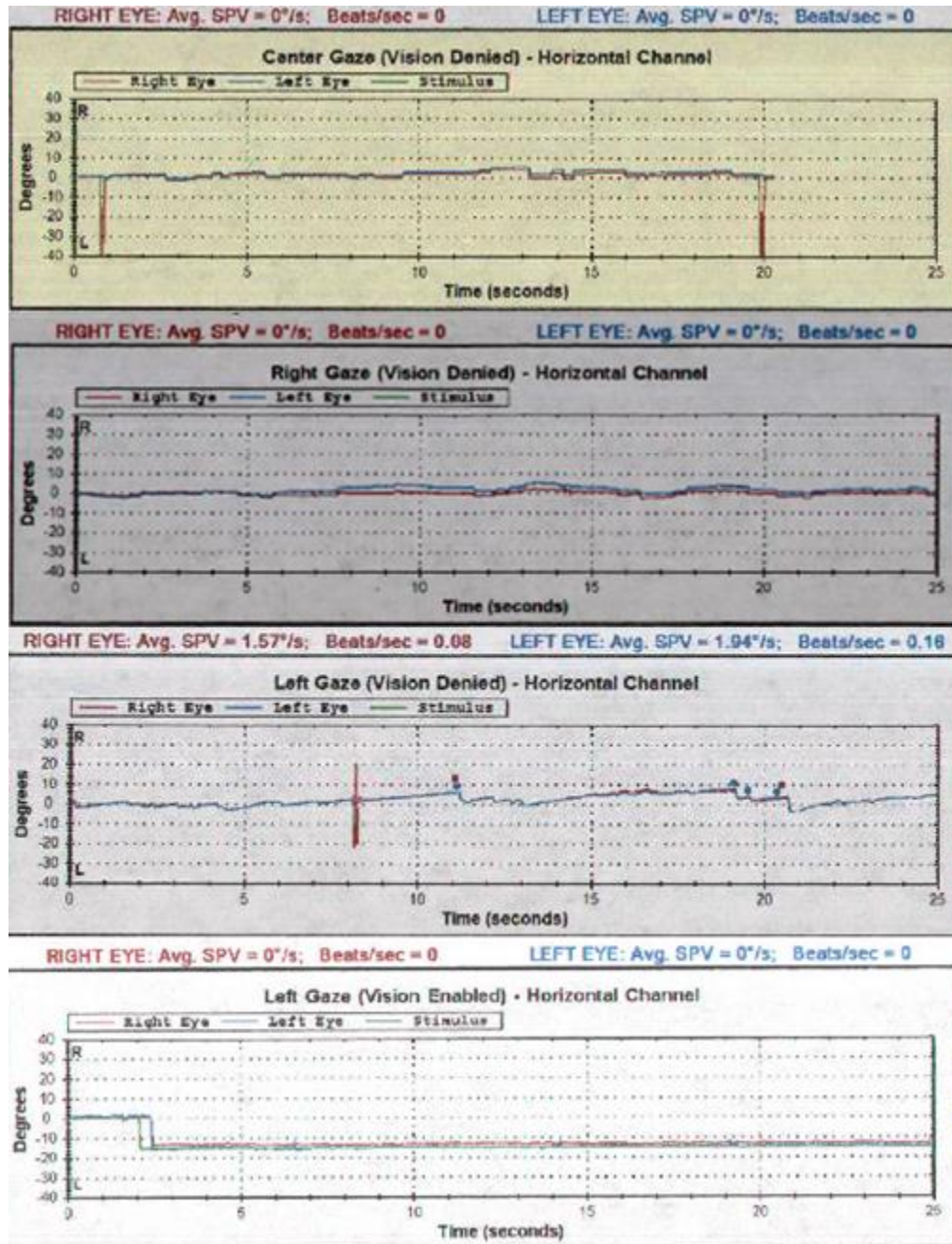


c. VNG- Oculomotor Evaluation



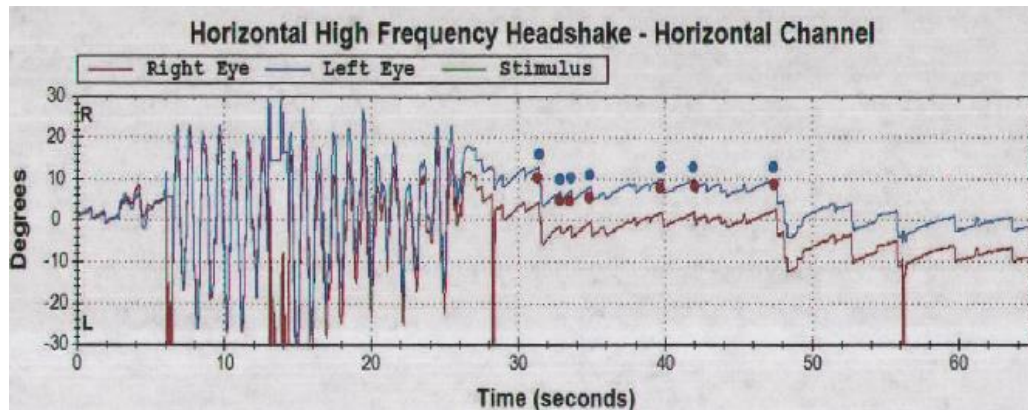
Description:
Normal saccades and OPKs. Corrective saccades were noted during smooth pursuit, suggestive of a peripheral vestibulopathy.

d. Gaze testing: Vision denied and vision enabled

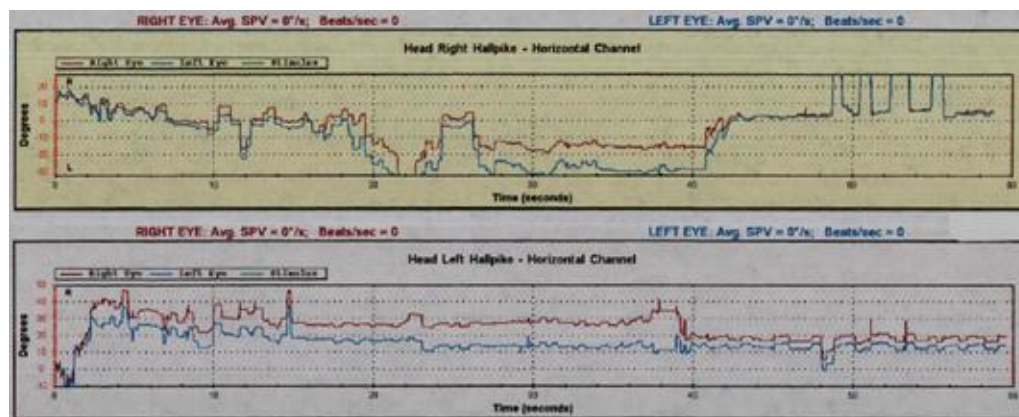


Description: LBN was noted during gaze left. Nystagmus suppressed with fixation

e. VNG: High frequency headshake and Dix-Hallpike

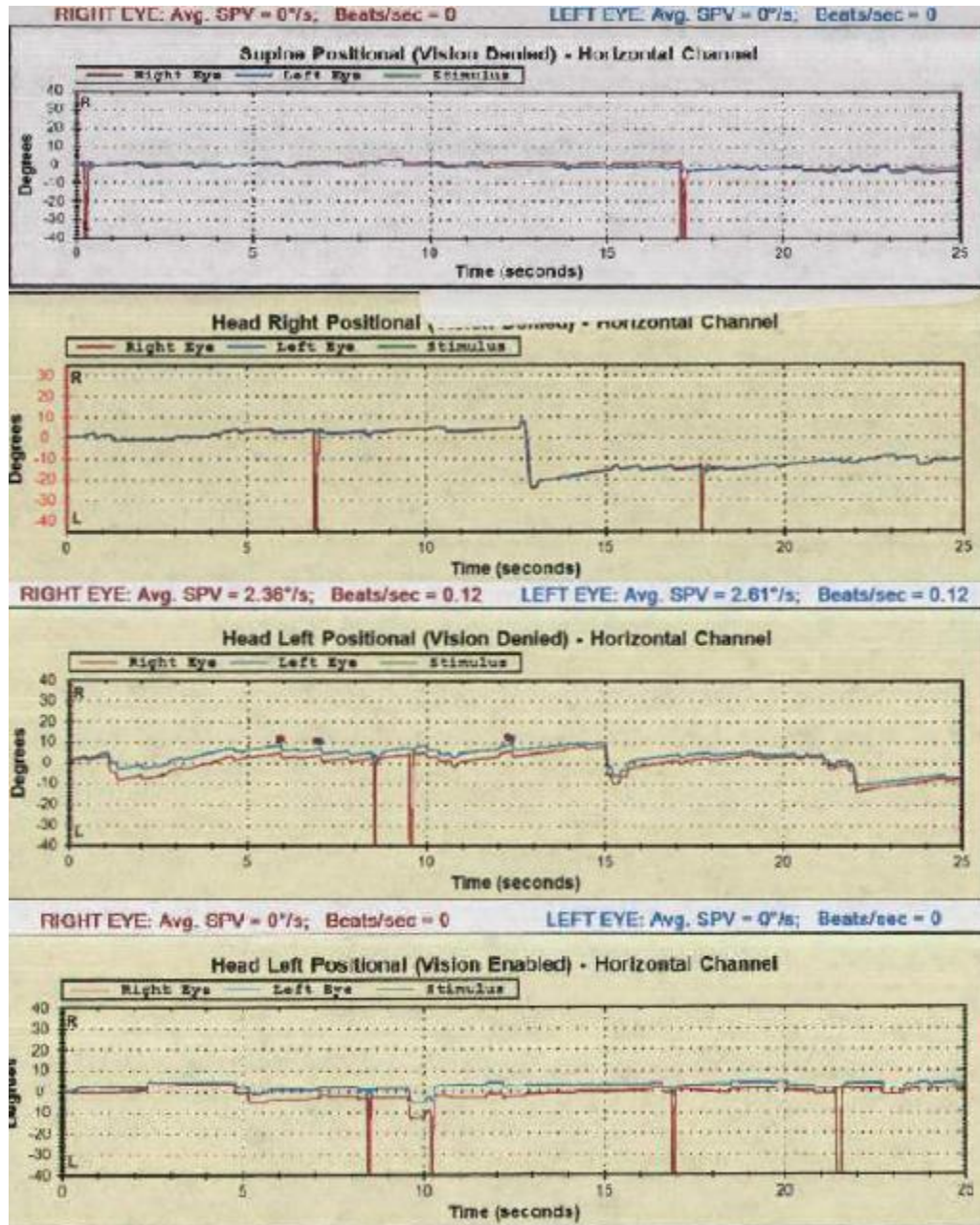


Description: LBN was noted post high-frequency headshake.



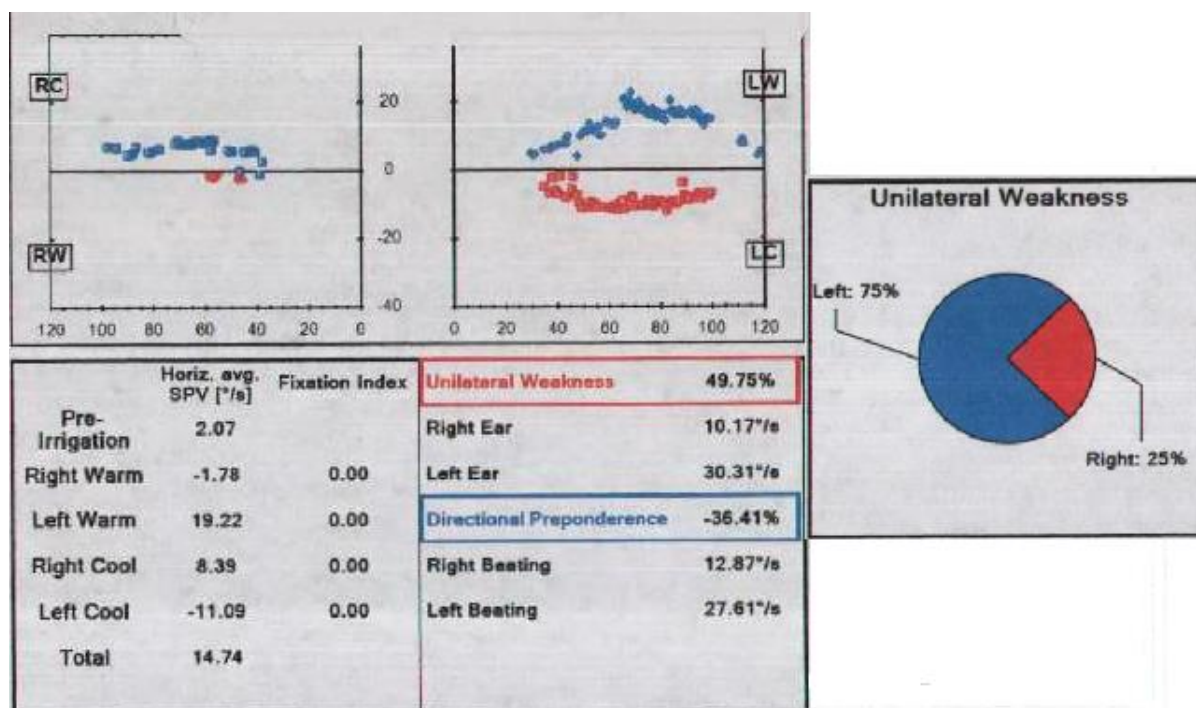
Description: Dix-Hallpike was unremarkable, bilaterally.

f. VNG- Positional evaluations, vision denied and vision enabled



Description: LBN was noted in the head left position. Nystagmus suppressed with fixation.

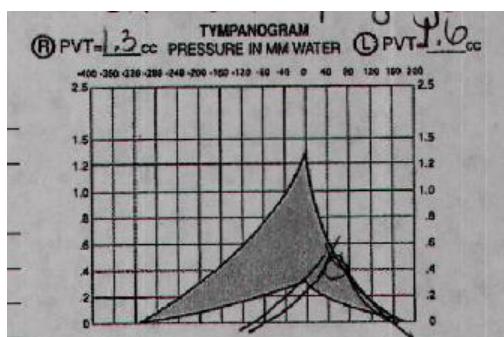
g. VNG- Bithermal Caloric Evaluation



Description: Bithermal caloric evaluation revealed a 50% unilateral weakness to the right.

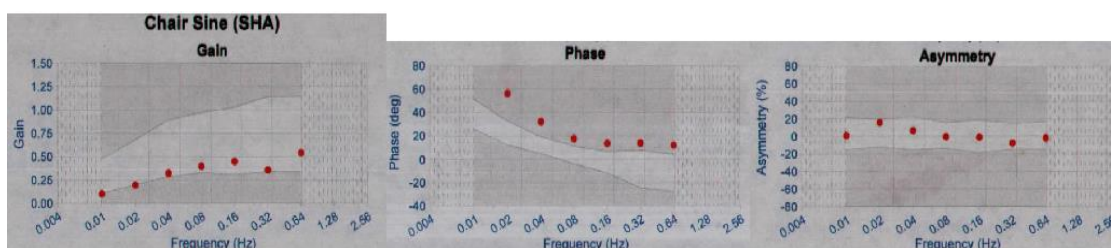
4. Case Number Three: S.M.

a. Tympanometry



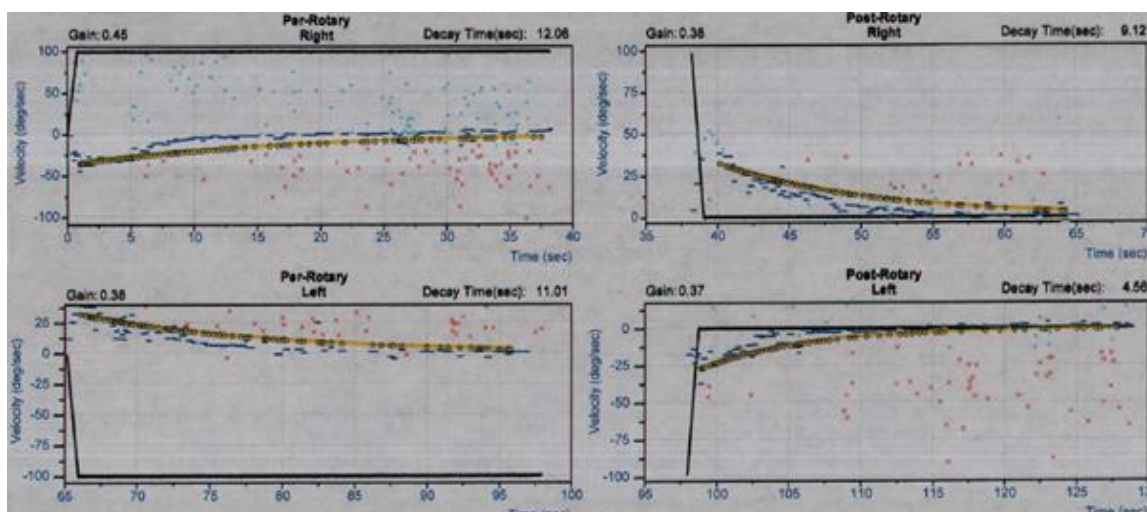
Description: Normal ear canal volume, compliance, and middle ear pressure, bilaterally. This suggests a normal middle ear system for bithermal caloric evaluation, bilaterally.

b. Rotational Chair- SHA



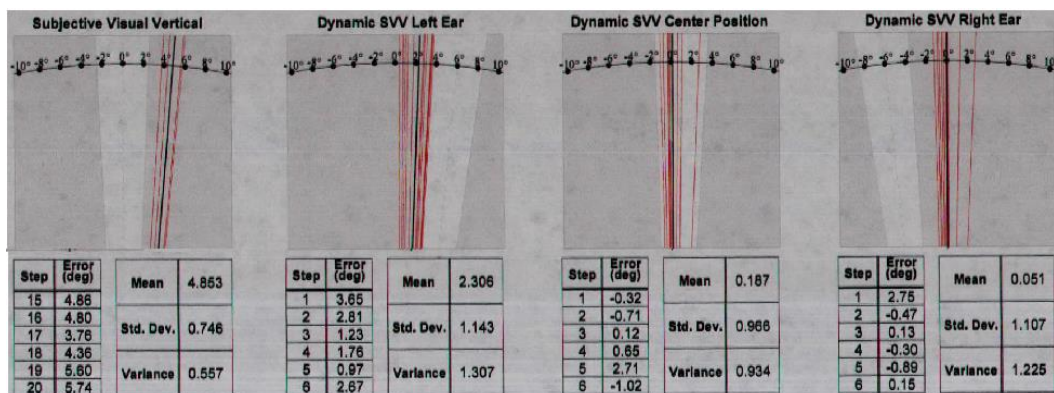
Description: Gain is reduced overall. Phase lead. This is consistent with a peripheral vestibular lesion. Normal symmetry was noted.

c. Rotational Chair- TRAPs



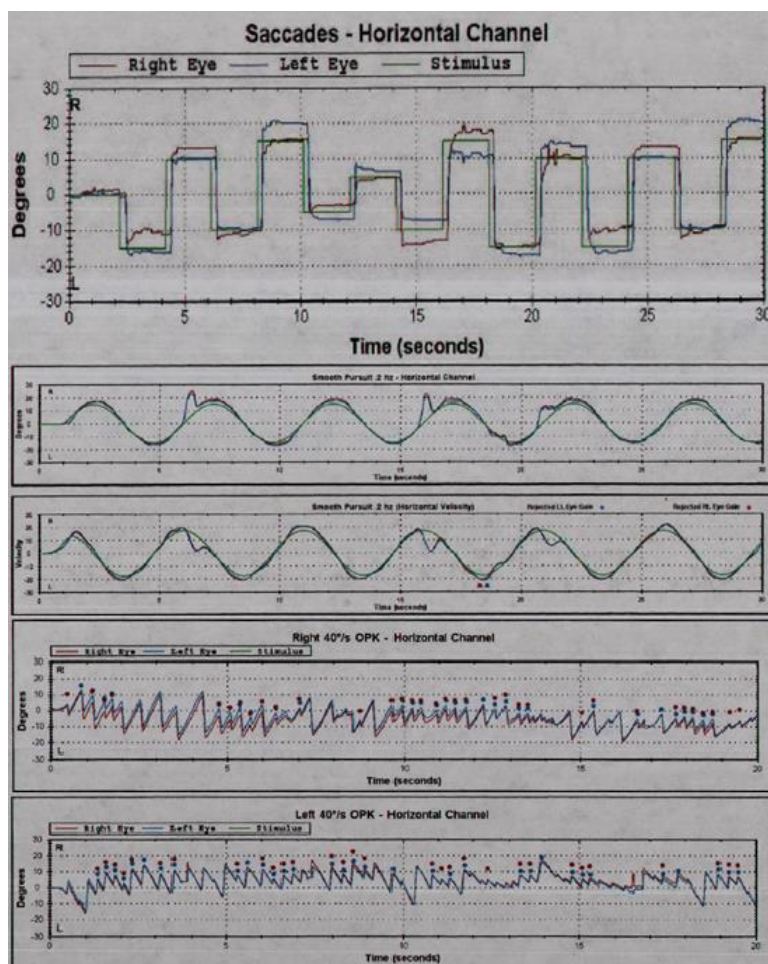
Description: Time constants are reduced overall, consistent with a peripheral vestibular lesion.

d. Rotational Chair- Static and dynamic SVV



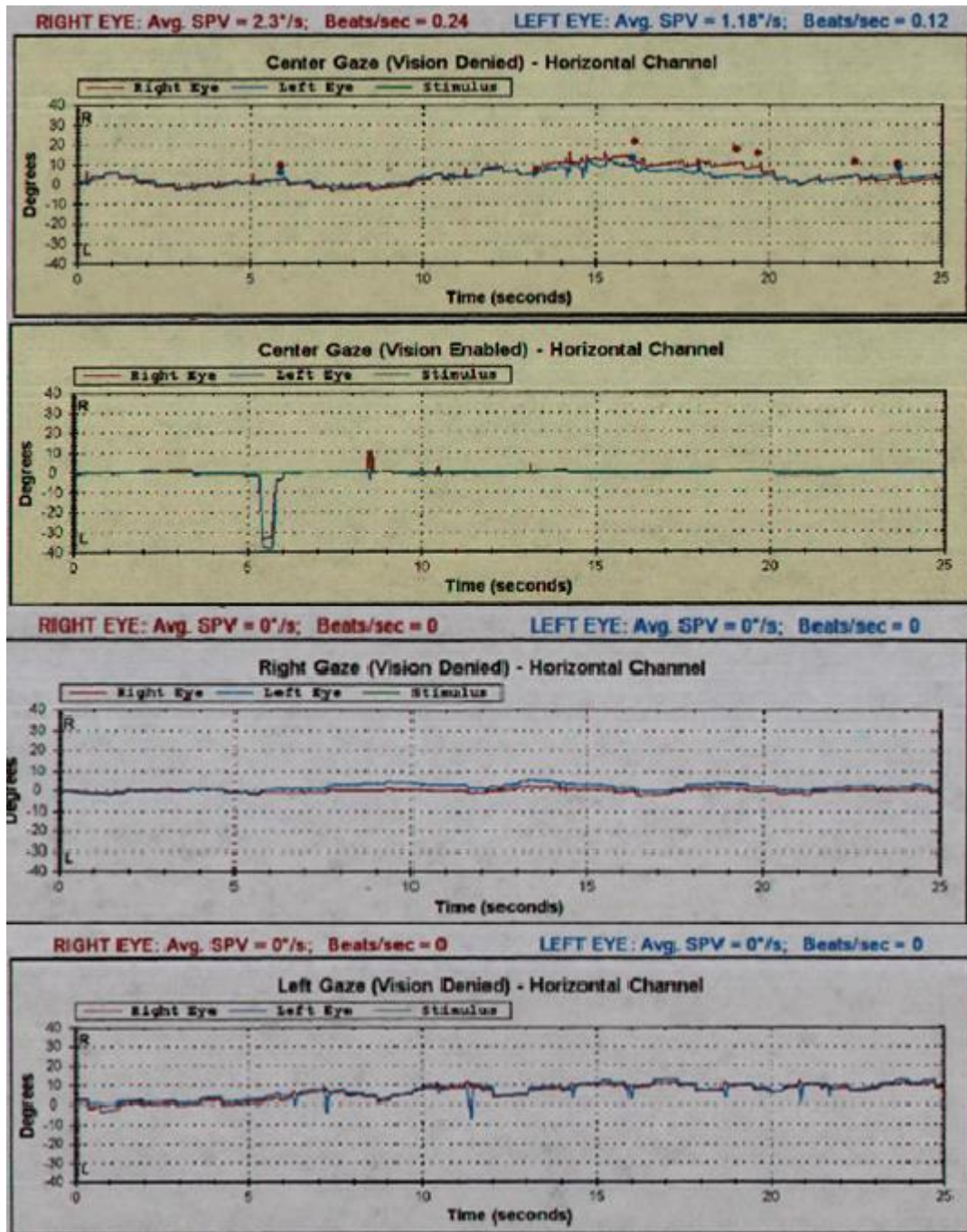
Description: Rightward preference in static and dynamic conditions, suggestive of uncompensated utricle dysfunction.

e. VNG.- Oculomotor Evaluation



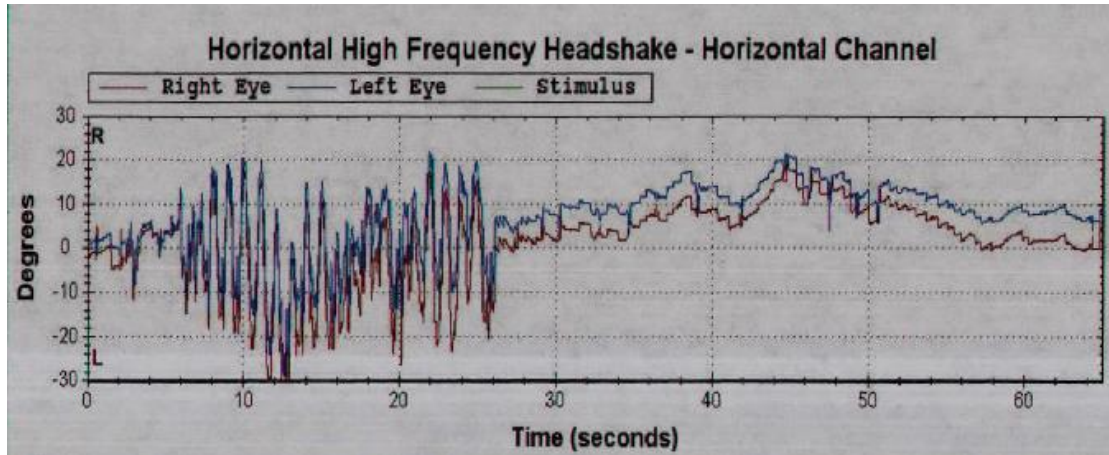
Description: Oculomotor testing is unremarkable.

f. VNG- Gaze testing, vision denied and vision enabled.



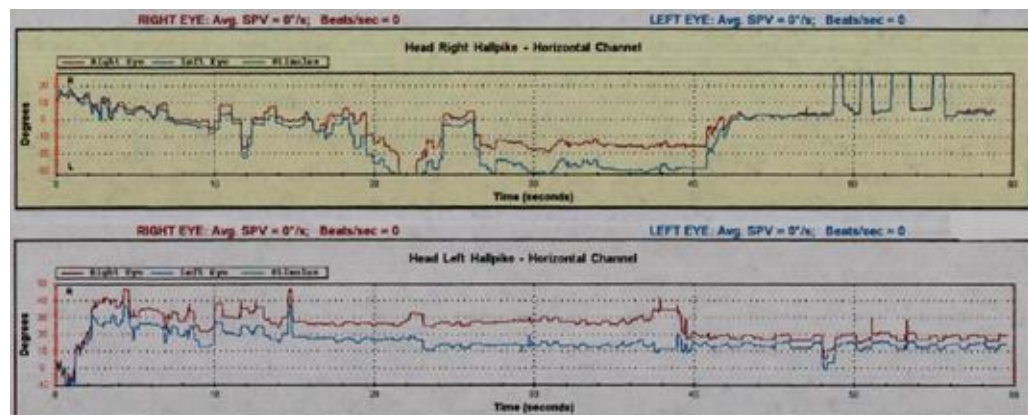
Description: Nystagmus was noted throughout testing; however nystagmus was no greater than spontaneous nystagmus. Nystagmus suppressed with fixation.

g. VNG- High frequency head shake and Dix-Hallpike



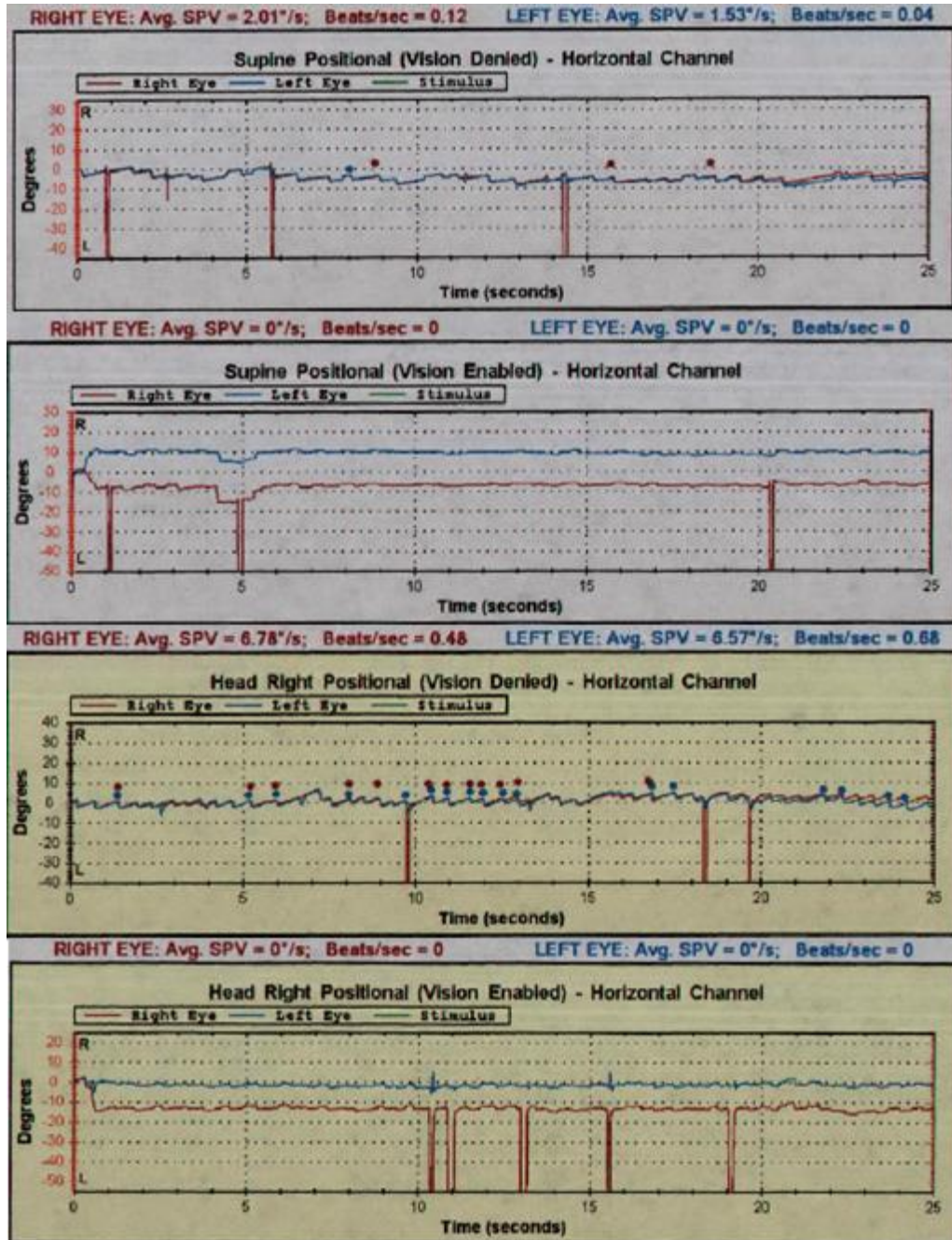
Description: RBN was visualized immediately post-headshake. After the RBN fatigued, spontaneous LBN was noted.

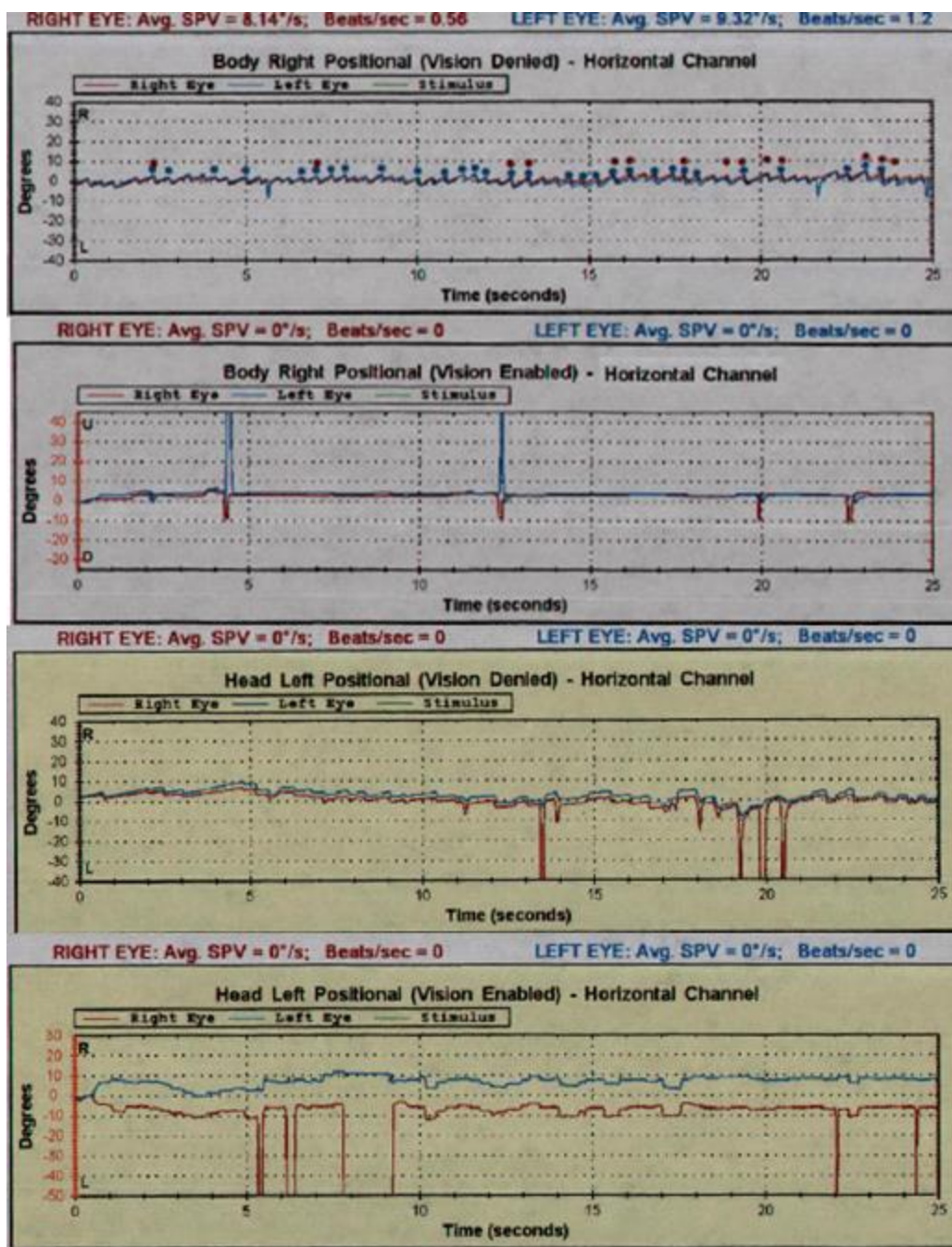
Dix-Hallpike



Description: Dix-Hallpike was unremarkable, bilaterally.

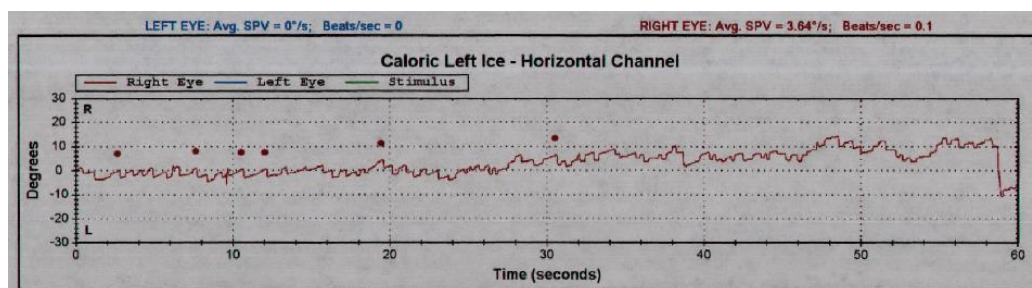
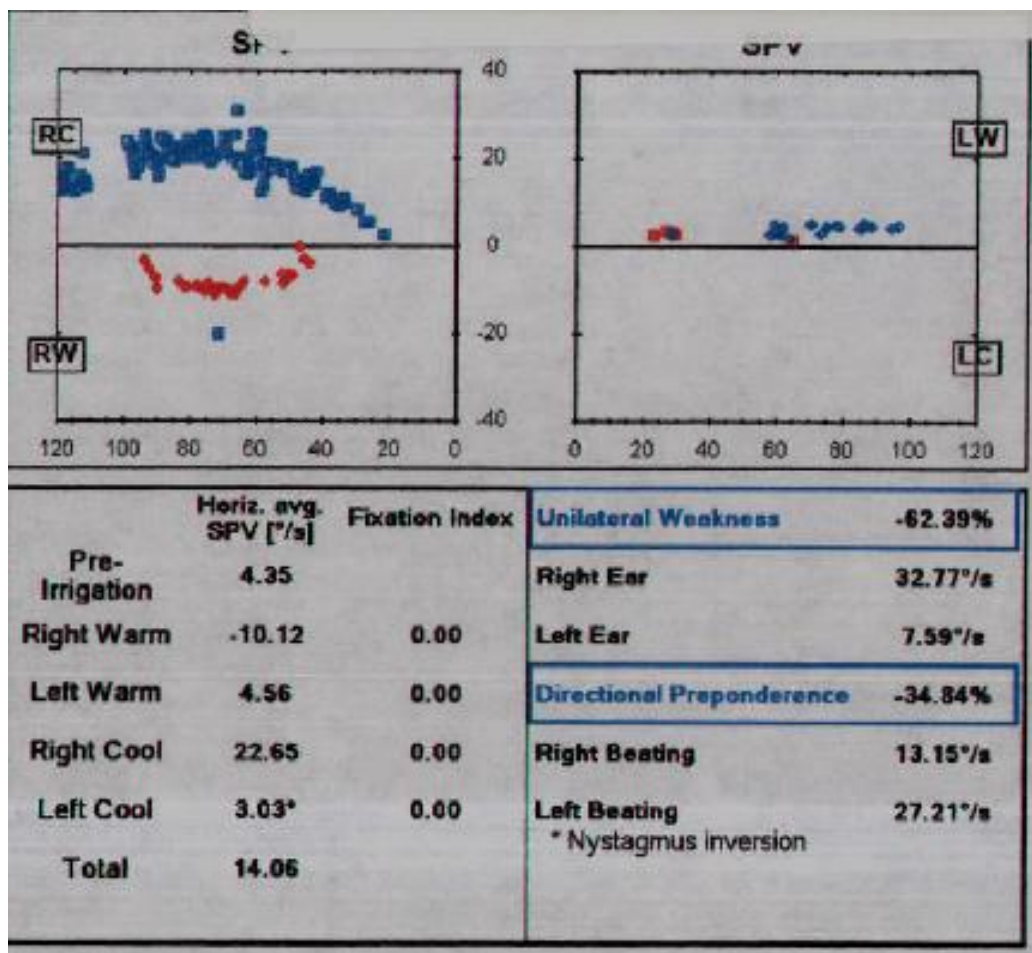
h. VNG- Positional evaluations





Description: Spontaneous LBN was noted in most positions, however all nystagmus suppressed with fixation.

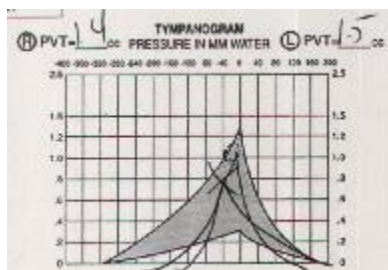
i. VNG- Bithermal caloric evaluation



Description: Reduced caloric responses for the right ear. No caloric responses for the left ear (nystagmus noted is spontaneous nystagmus). A unilateral weakness of 62% to the left was noted. Ice water calorics were performed in the left ear because there was no response from the left ear. The response from ice water calorics is no greater than spontaneous nystagmus, which suggests left canal paralysis.

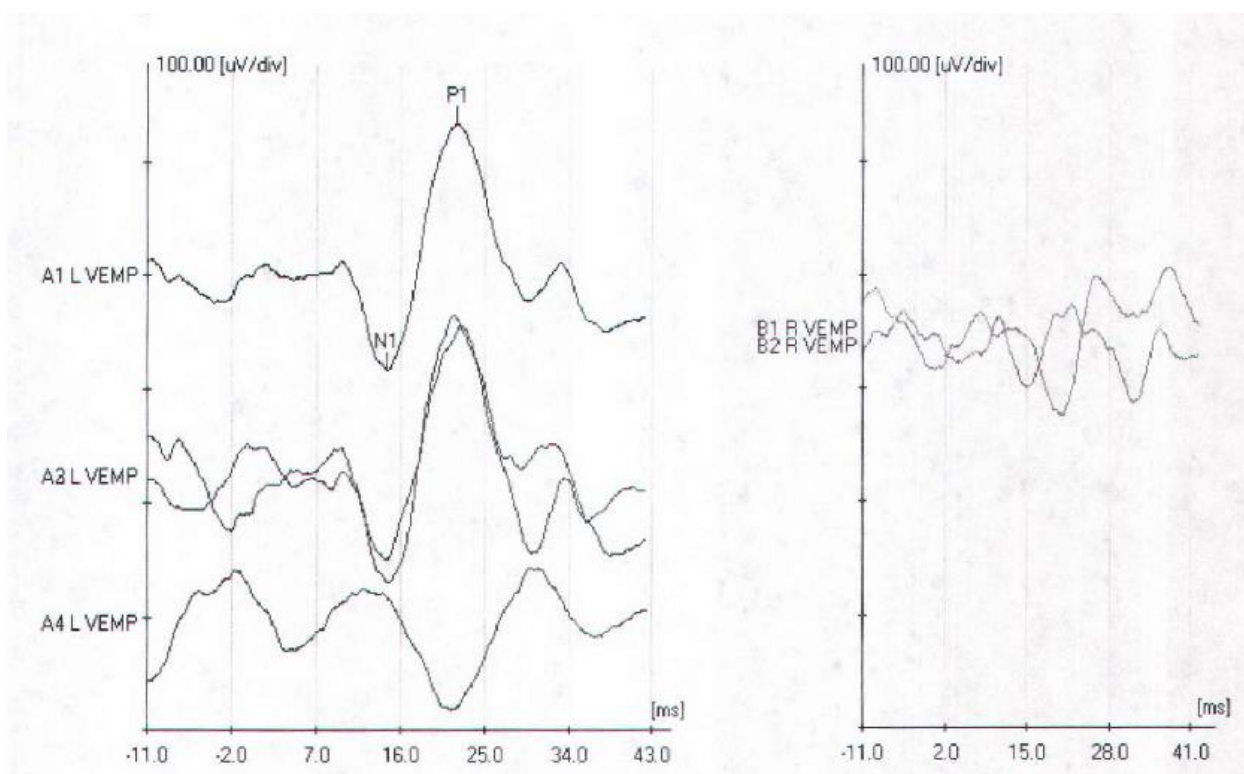
5. Case Number Four: S.R.

a. Tympanometry



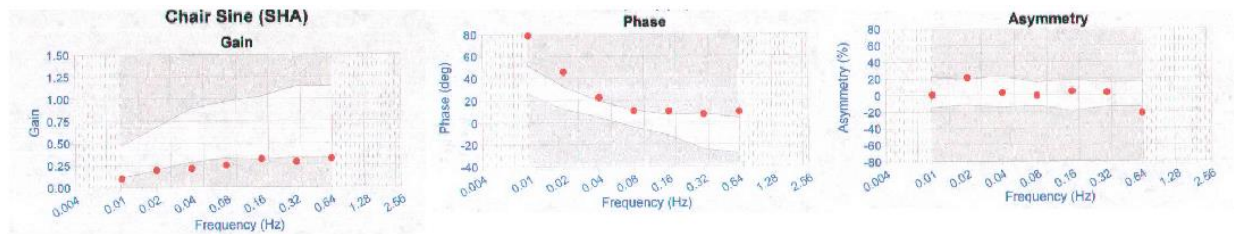
Description: Normal ear canal volume, compliance, and middle ear pressure, bilaterally. This suggests a normal middle ear system for bithermal caloric evaluation, bilaterally.

b. C-VEMP



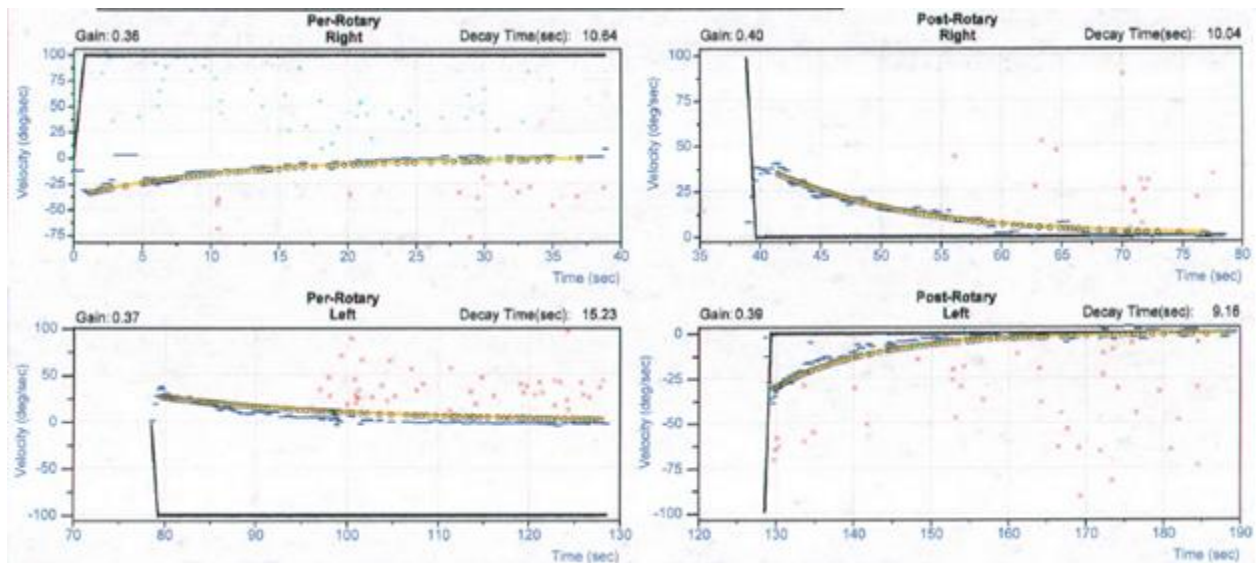
Description: Absent response on the right and normal response on the left. Asymmetry ratio: 100%. This is consistent with diagnosis of right acoustic neuroma.

c. Rotational Chair- SHA



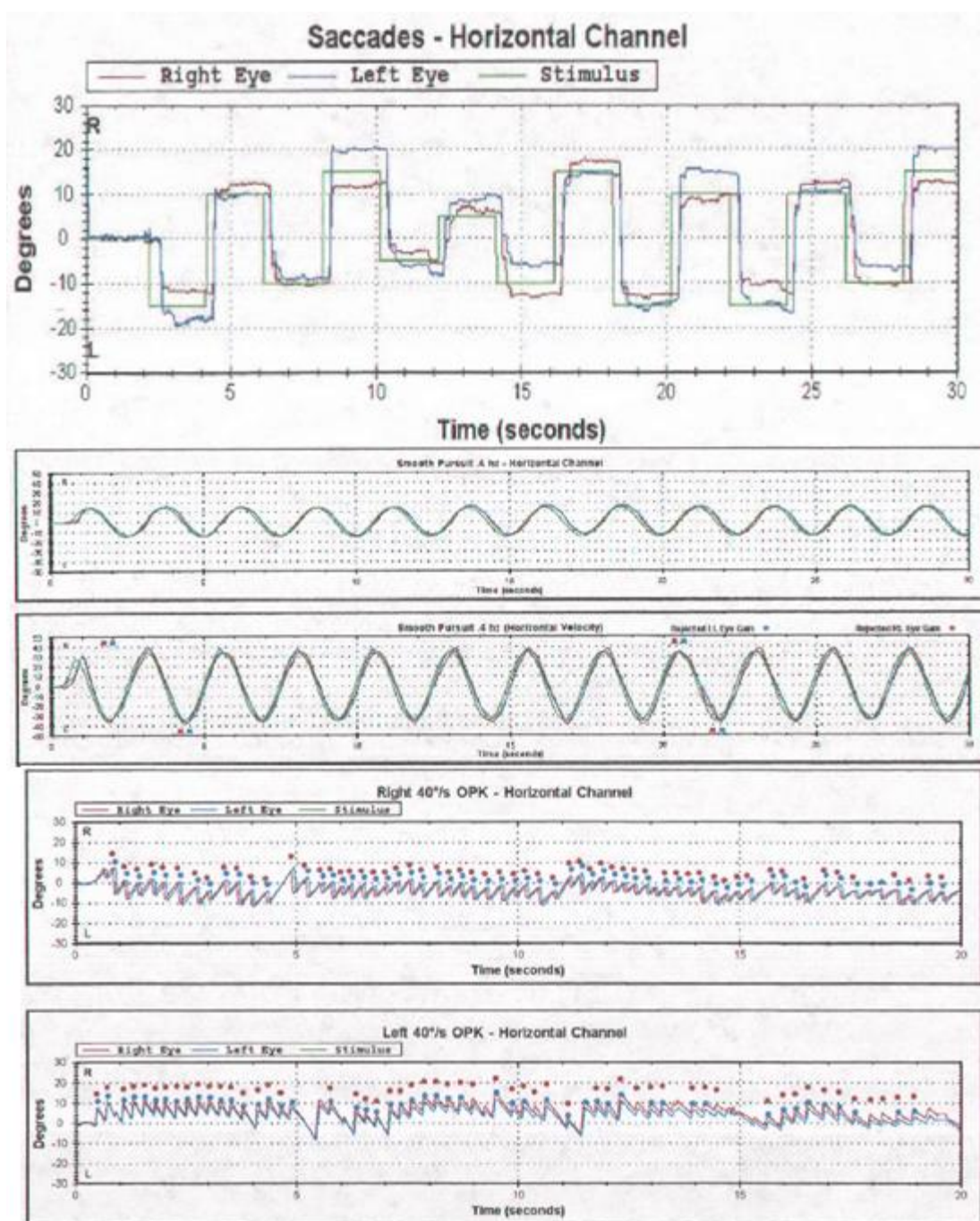
Description: Reduced gain and phase lead, which is consistent with a peripheral vestibulopathy and patient diagnosis of unilateral acoustic neuroma.

d. Rotational Chair- TRAPs



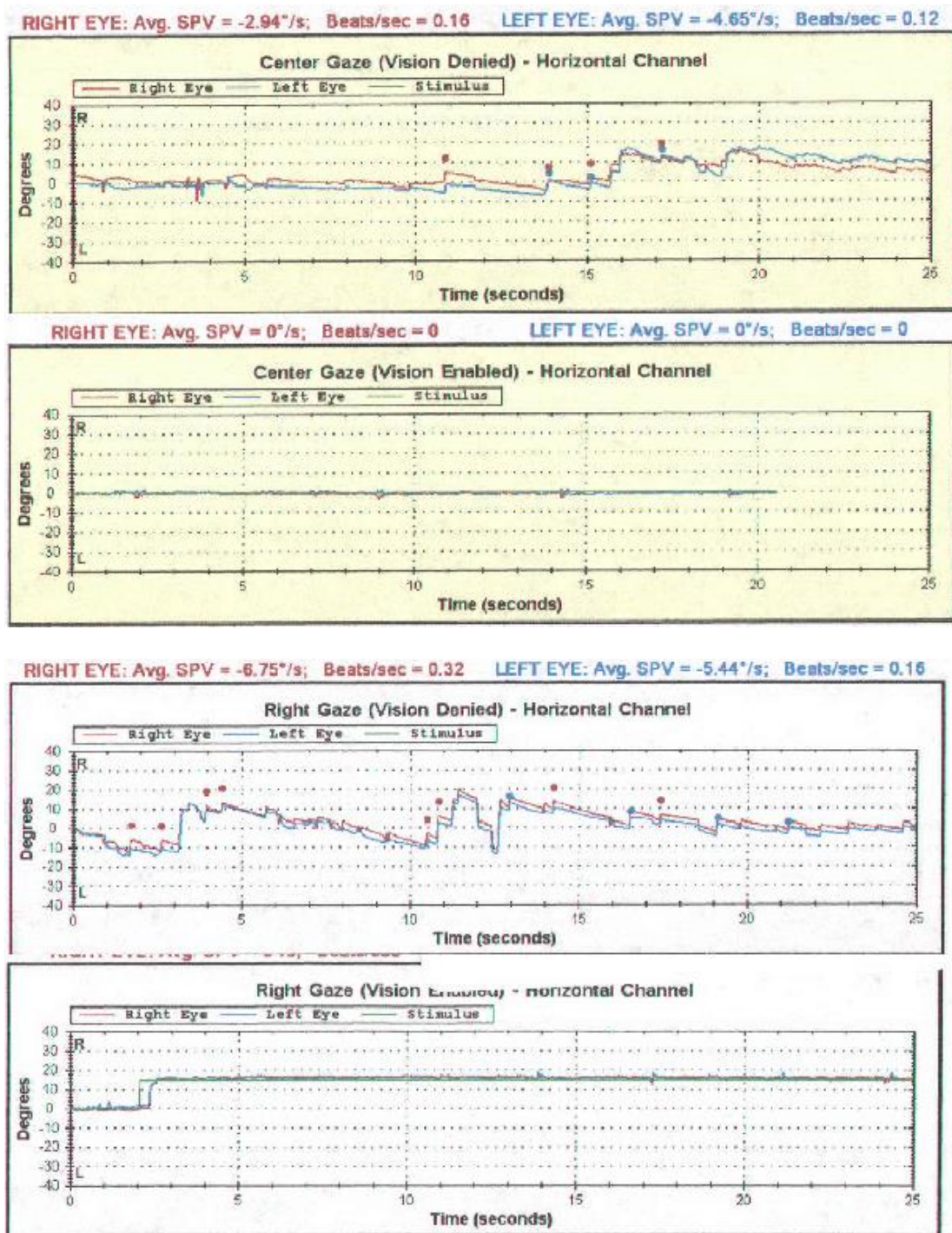
Description: Time constants are essentially reduced, which is consistent with a peripheral vestibulopathy and patient diagnosis of unilateral acoustic neuroma.

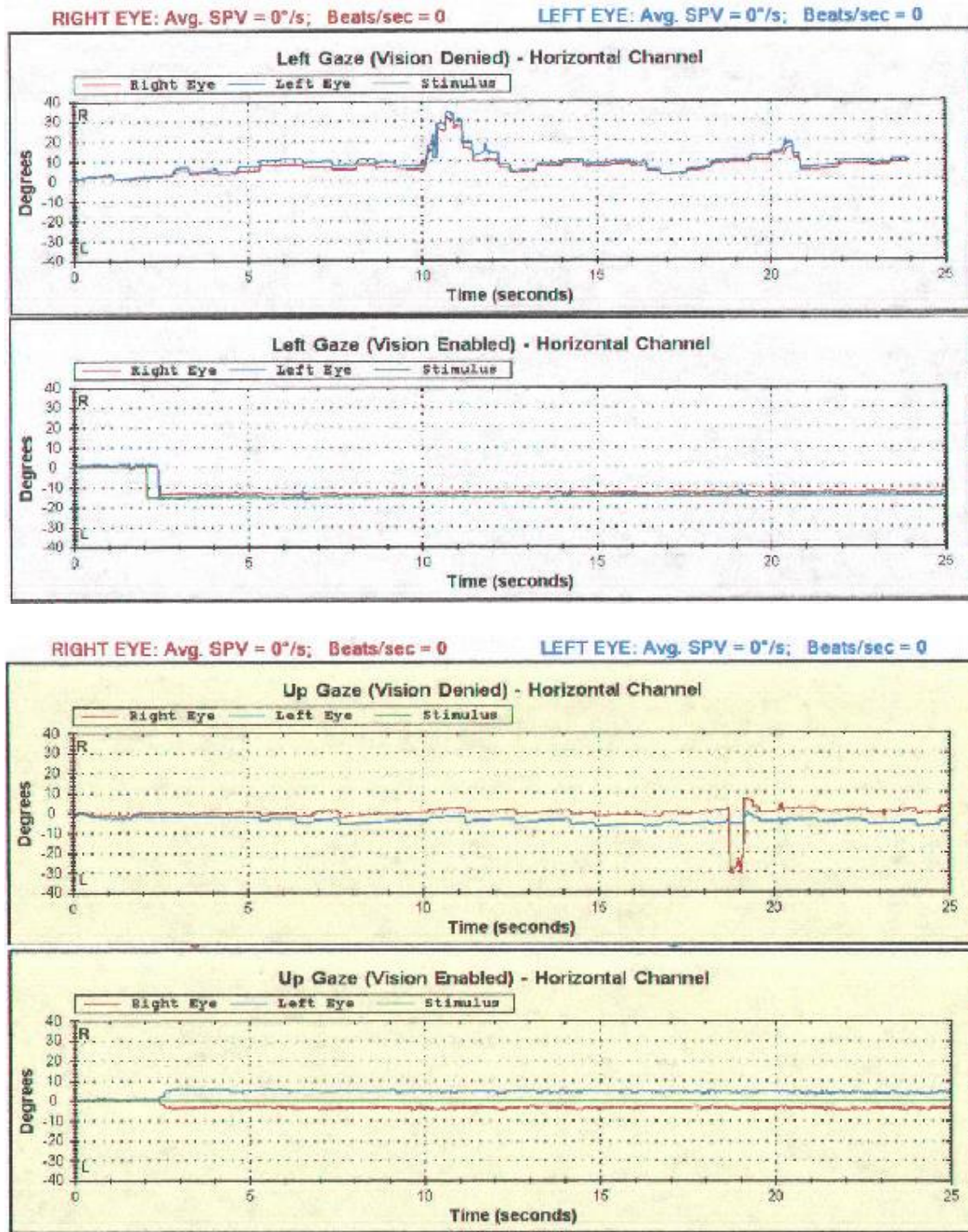
e. VNG- Oculomotor testing



Description: Oculomotor testing is unremarkable.

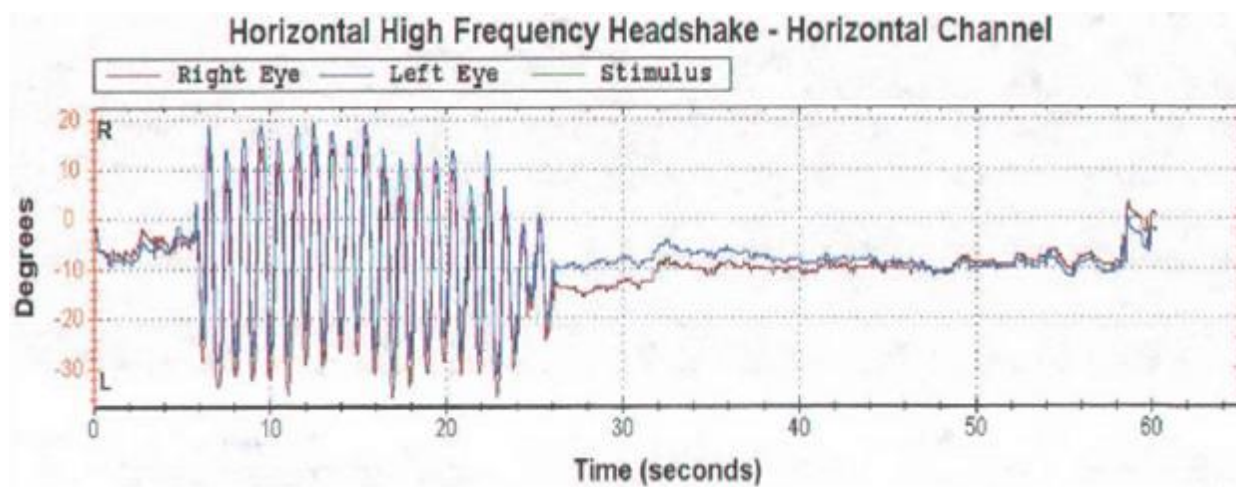
f. VNG- Gaze- vision denied and vision enabled



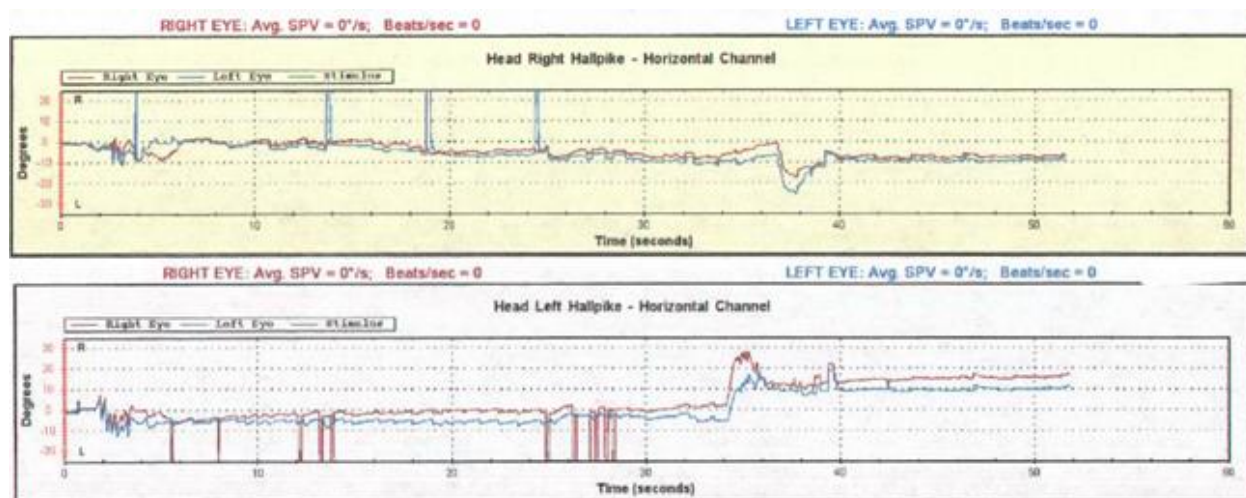


Description: RBN was noted in all conditions with vision denied. Nystagmus suppressed with fixation.

g. VNG- High frequency head shake and Dix-Hallpike.

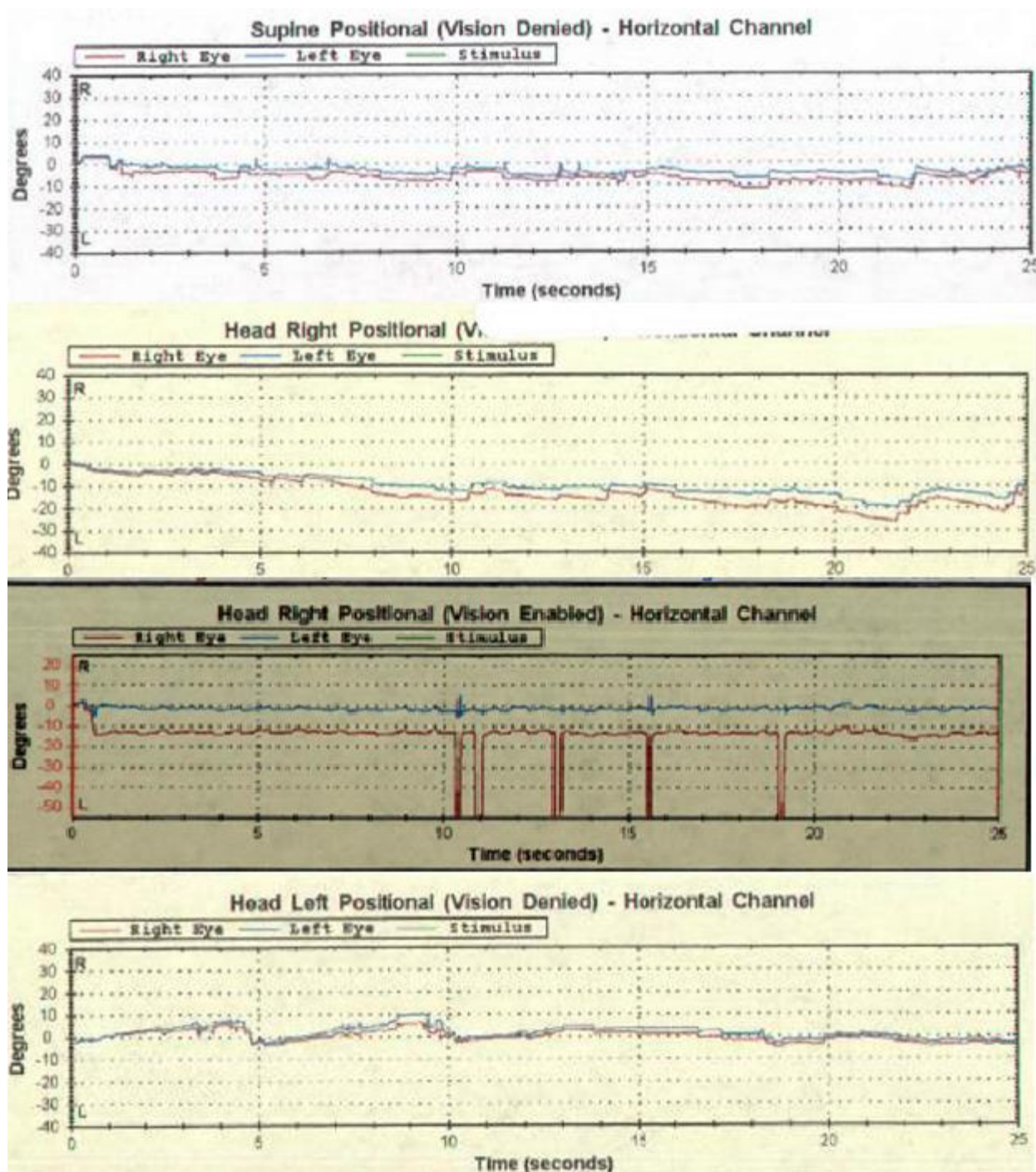


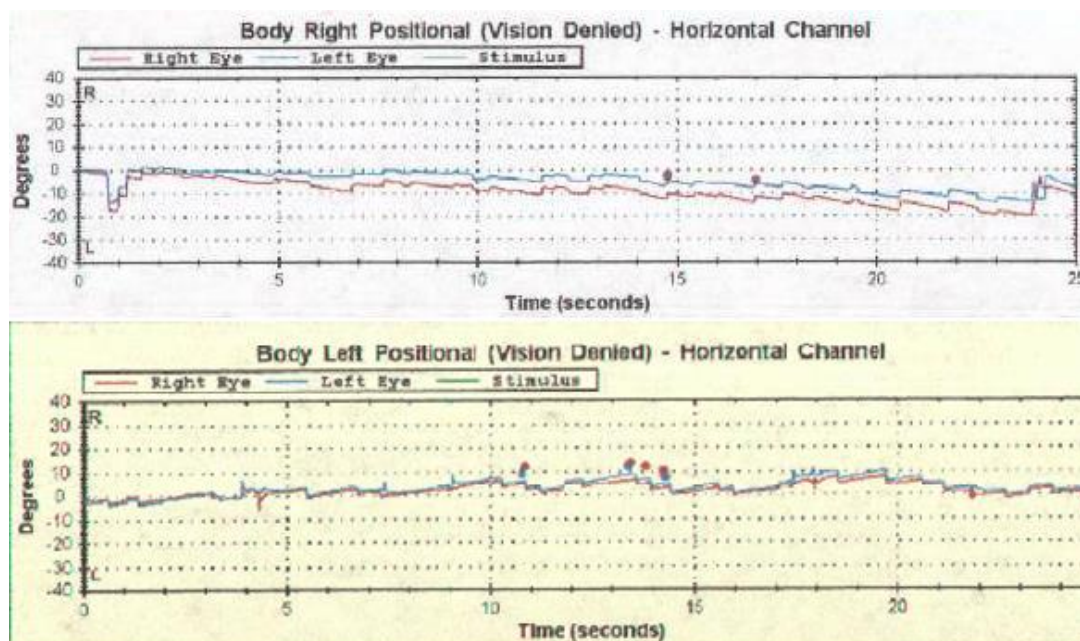
Description: Minimal LBN was visualized immediately post headshake.



Description: Dix-Hallpike was unremarkable, bilaterally.

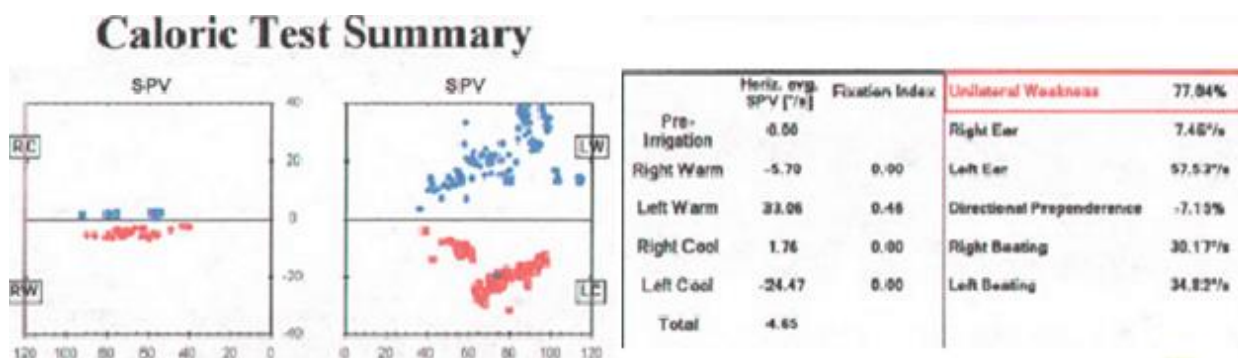
h. VNG- Positional testing- vision denied and vision enabled





Description: Minimal geotropic nystagmus was noted in all positions during positional evaluations and nystagmus suppressed with fixation in all positions (results of vision enabled are not shown above). This is consistent with patient diagnosis.

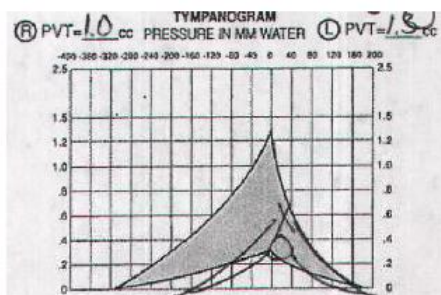
i. VNG- Bithermal Caloric Evaluation



Description: Bithermal caloric responses revealed a 77% unilateral weakness to the right, consistent with patient diagnosis of right acoustic neuroma.

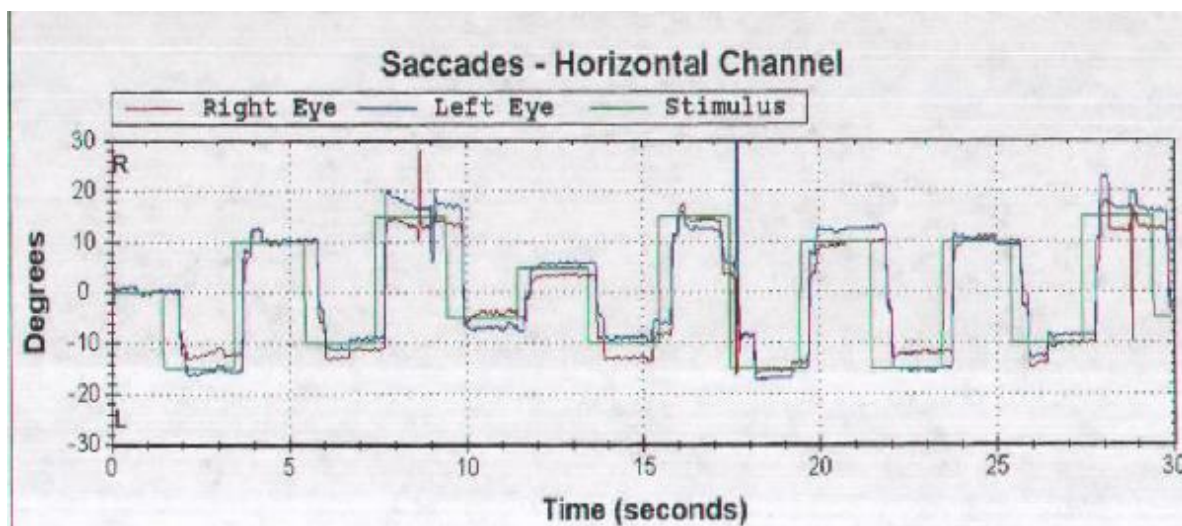
6. Case Number Five: P.P.

a. Tympanometry

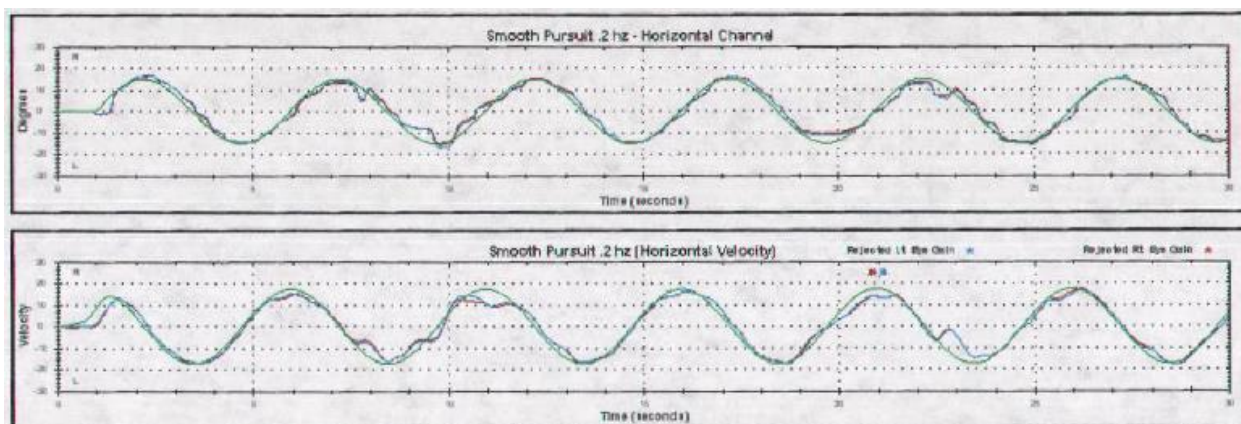


Description: Normal ear canal volume, compliance, and middle ear pressure, bilaterally. This suggests a normal middle ear system for bithermal caloric evaluation, bilaterally.

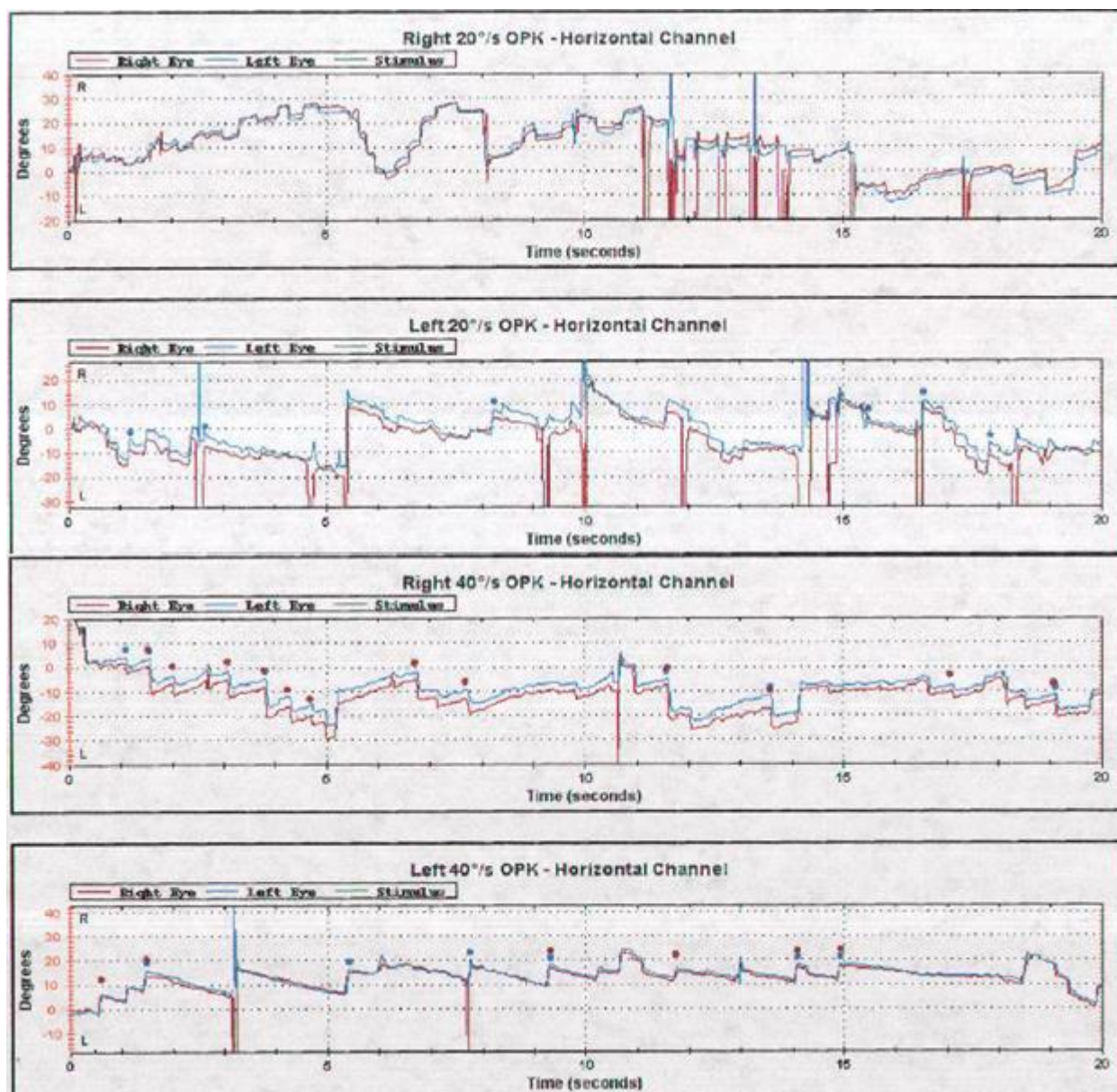
b. VNG: Oculomotor Testing



Description: Increased latencies and poor accuracy noted.

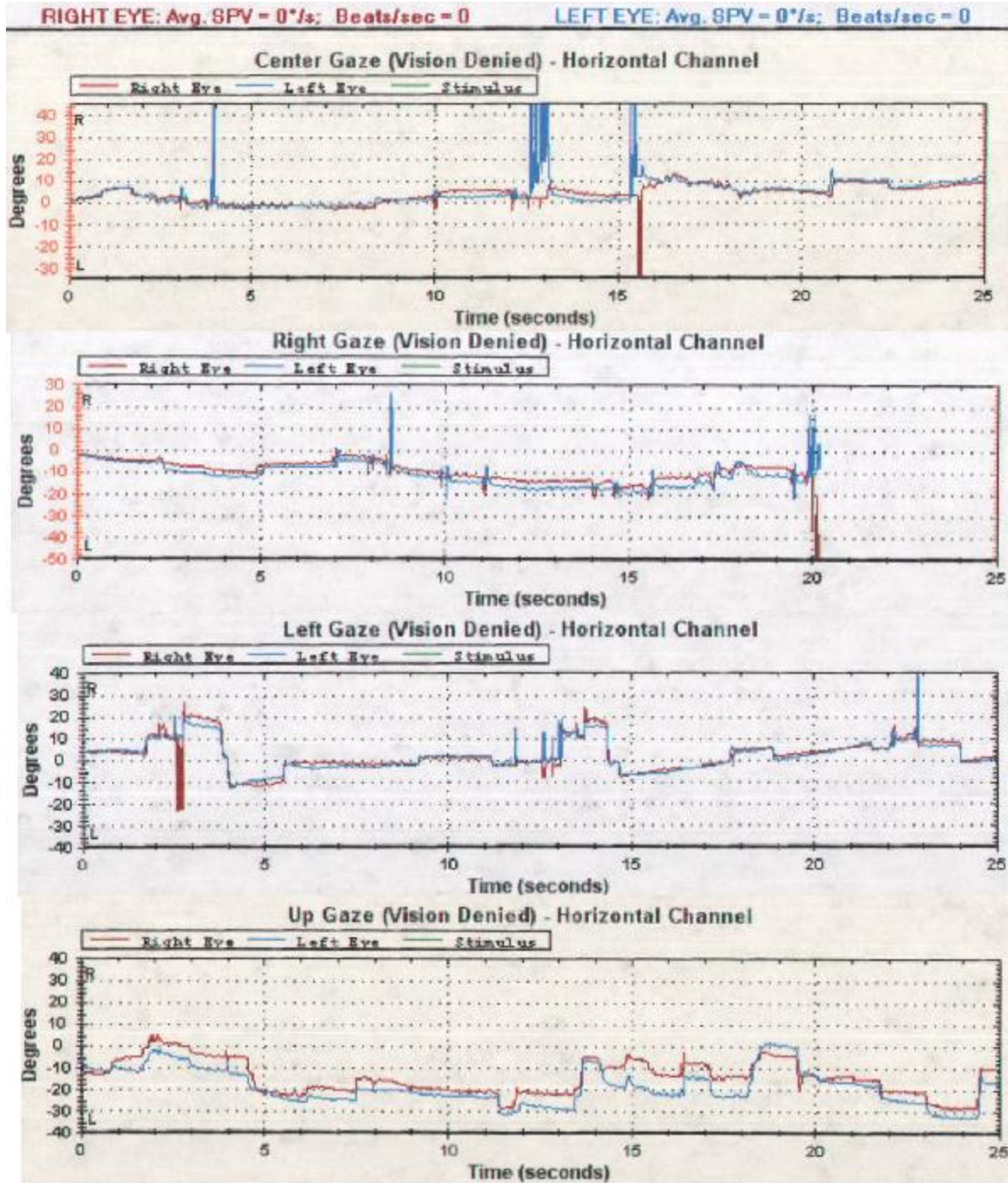


Description: Saccadic catch-ups noted during smooth pursuit.



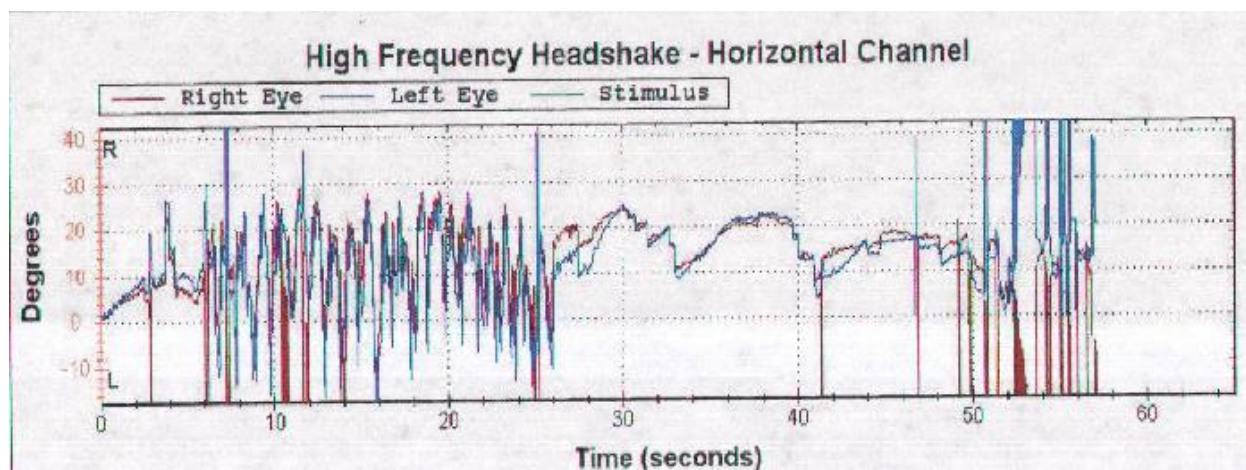
Description: Significantly decreased gain was noted in all conditions.

c. VNG: Gaze testing, vision denied

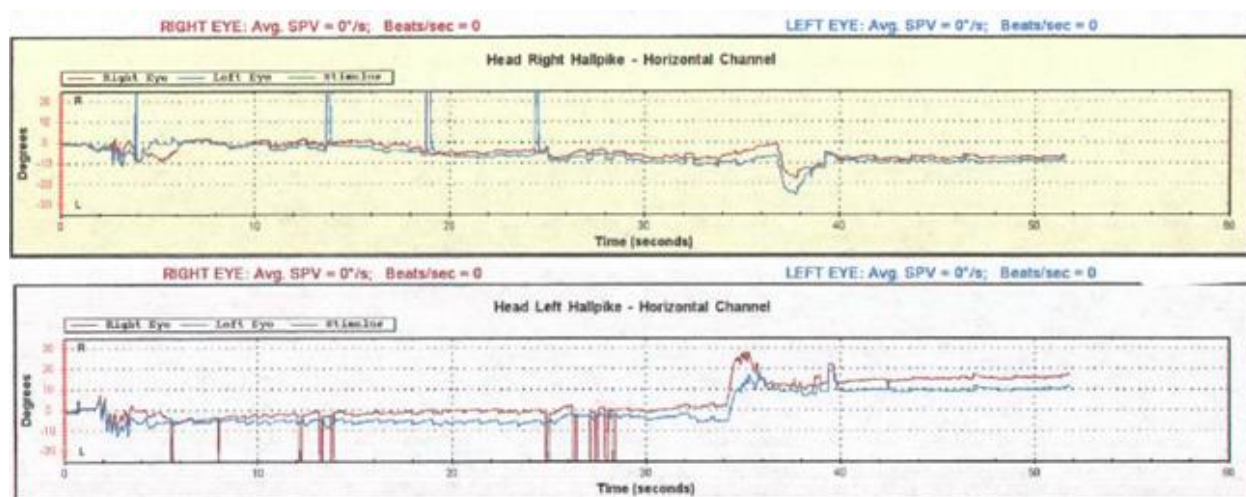


Description: Gaze testing was unremarkable.

d. VNG: High-frequency headshake and Dix-Hallpike

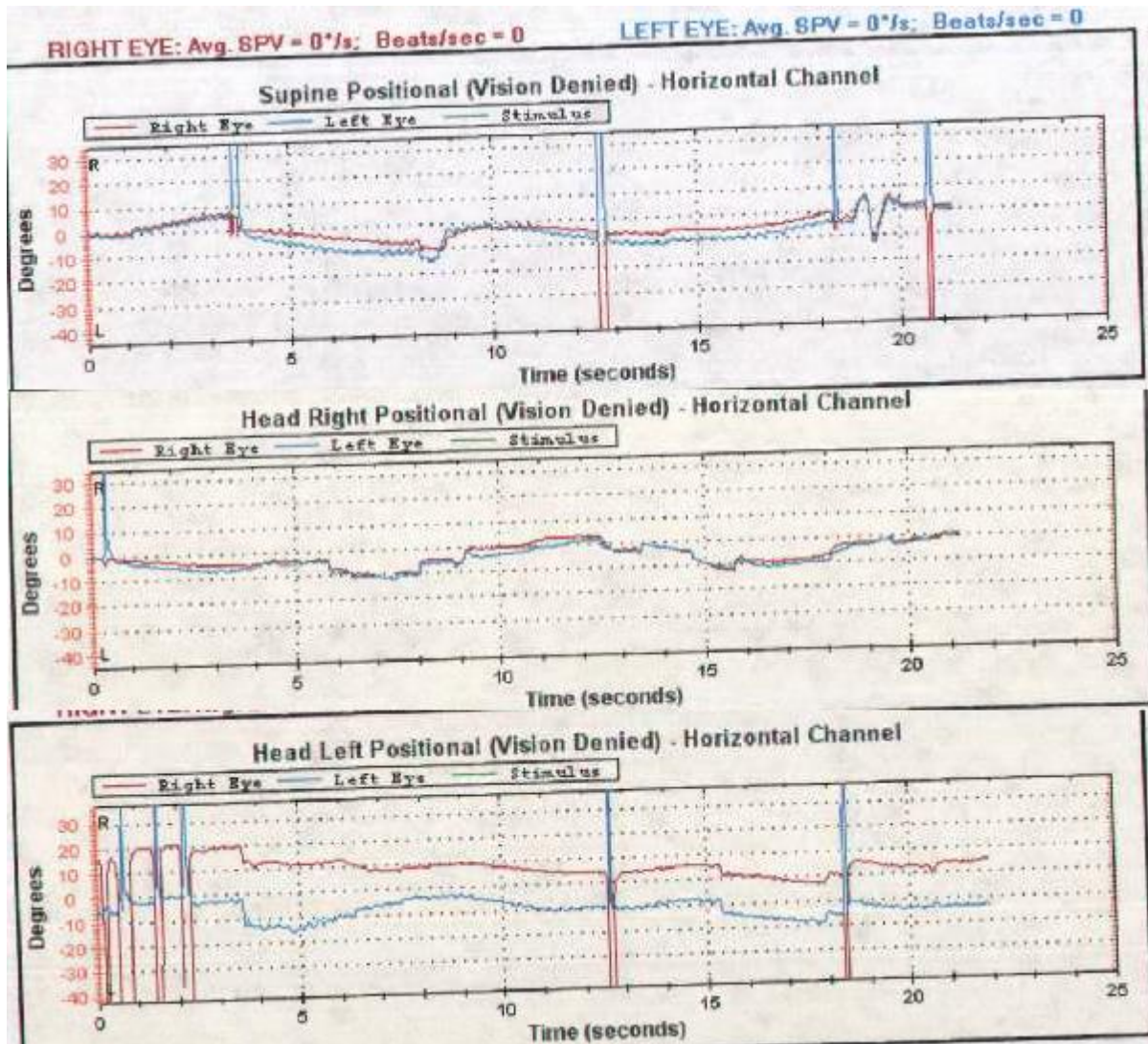


Description: Unremarkable



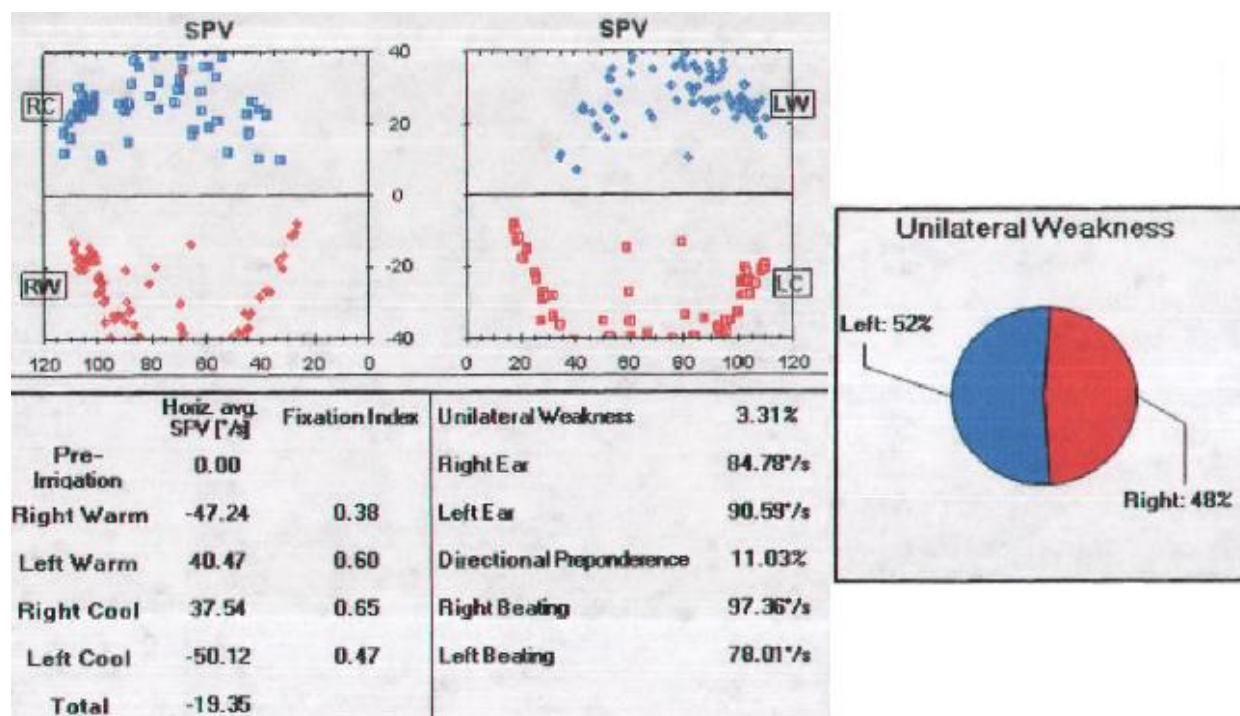
Description: Unremarkable

e. VNG: Positional tests



Description: Unremarkable

f. VNG: Bithermal Caloric Evaluation



Description: Hyperactive responses, bilaterally. Responses are symmetric. An increased fixation index was noted in multiple conditions.

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